STUDY OF CONGENITAL HEART DISEASE IN BUNDELKHAND REGION

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C E R T I F I C A T E

This is to certify that the work entitled "STUDY OF CONGENITAL HEART DISEASES IN BUNDELKHAND REGION" which is being submitted as thesis for M.D. (Pediatrics) examination, 1991 of Bundelkhand University by POONAM ATRI has been carried out in the department of Pediatrics.

She has put in necessary stay in the department according to University regulations.

Dated : December, 4, 1990

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C E R T I F I C A T E

This is to certify that the work entitled "STUDY OF CONGENITAL HEART DISEASE IN BUNDELKHAND REGION" which is being submitted as a thesis for M.D. (Pediatrics) by POONAM ATRI has been carried out under my supervision and guidance in the department of Pediatrics. The techniques embodied in the thesis were undertaken by the candidate herself and the observations recorded have been regularly checked by me.

She has fulfilled necessary requirements of the stay in the department for the submission.

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C E R T I F I C A T E

Certified that the work entitled "STUDY OF CONGENITAL HEART DISEASES IN BUNDELKHAND REGION", which is conducted by POONAM ATRI was carried out under my guidance, by the candidate herself.

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C O N T E N T S

INTRODUCTION

INTRODUCTION

Congenital heart disease is defined as abnormality at birth in structure or function of heart. They result generally from altered embryonic development of a normal structure or failure of such a structure to progress beyond an early stage of embryonic or fetal development. The aberrent pattern of flow by an anatomical defect may in turn, significantly influence the structural and functional development of the remainder of circulation. There are also certain congenital defects that are not apparent on gross inspection of heart or circulation. Examples include the electrophysiological path way for ventricular preexcitation or interruption in the cardiac conduction system giving rise to paroxysmal supreventicular tachycardia or congenital complete heart block, respectively.

Most congenital defects are well tolerated during fetal life. Only after the elimination of maternal circulation the abnormalities becomes apparent. The infant circulation continues to change after birth and weeks, months or even years may elapse before the anomaly evolves into the typical clinical picture. Both physiologic and structural changes subsequently continue, or conversely, the malformation may 'vanish'.

The ductus in premature infant sometimes remain widely patent for months, finally closing spontaneously, leaving the baby with a normal heart. A ventricular septal defect that delivers a large left to right shunt in infancy may gradually develop progressive infundibular pulmonary stenosis, so that years later the physiologic and the clinical picture resemble classic cyanotic Fallot's tetralogy. A congenital bicuspid aortic valve that is functionally normal at birth may take two or more decades to stiffen, calcify and present as overt aortic stenosis. Thus congenital heart disease should not be viewed narrowly as a fixed ground of anatomic defects present at birth but as a dynamic group of anomalies that originate in fetal life and after during postnatal development.

On the other hand with the changes occuring in hemodynamics, there occurs changes in cardiac lesion for example as pulmonary vascular resistance falls over the first week of life, left to right shunt become more apparent. The relative significance of various defects also changes with growth as many of them become smaller later on (V.S.D.) or some become worse (Aortic or pulmonary stenosis).

The importance of congenital heart disease in pediatric cardiology can scarcely be over emphasized. Until the last three decades rheumatic heart disease was believed to be common form of cardiac disease in children. Later, it has become obvious that congenital heart disease is more common of the two. The shift of emphasis towards congenital heart disease in children is the result of spectacular advances in surgical treatment of congenital heart disease and downgrade trend of rheumatic fever.

Various surveys carried out in developed countries report incidence of congenital heart disease varying from 0.75% to 1.17% in live births (Kerrebijin et al, 1966; Mithell et al, 1971; Yerushalury, 1976; Rose et al, 1964). The incidence is high in premature and still births them in full term births. Among infants born with cardiac defects there is a spectrum of severity, about 20-25% infants with congenital heart disease will be symptomatic in the first year of life. Since palliative and corrective surgical techniques have evolved, the percentage of individuals who survive with various lesions has changed over the years, complex severe defects later in childhood now account for a large number of patients.

The pattern of anomalies is different in India than those from developed countries. V.S.D. is most common anomaly, seen in USA & Canada, while studies from U.K. & Europe show A.S.D. & P.D.A. to be more common lesion. The different figures obtained depending on whether one discusses living patients or results of post mortem examinations, in addition each author's series varies in terms of age or type of disease.

In toto, children with congenital heart disease are predominantly male. Moreover, specific defect may show a definitive sex predonderence. PDA & ASD are more common in females, where as valvular aortic stenosis, congenital aneurysm of sinus of valsalva, coarctation of aorta, tetralogy of Fallot's & T.G.A. are more common in males.

The etiology of congenital heart disease is unknown in most instances. A multifactorial inheritance hypothesis is gaining increasing acceptance, rarely single gene syndromes, gross chromosomal abnormalities may found to be responsible (3% and 5% cases respectively). In most instances there is a combination of genetic and environmental influences.

In 2% cases environmental factors and in 90% cases environmental factors with associated genetic factors are responsible for causation of disease.

Extracardiac anomalies occur in approximately 25% of infants with significant cardiac disease, one third of them have some established syndrome as Turner Noonan's, Leopard, Holt-oram, Ellis-van-crevald, Xerta-gener, Laurence - Moon-Biedl, Marfan syndrome etc.

There is approximately a 1% incidence of congenital heart disease in normal population and this incidence increases to 2-6% for a second pregnancy following the birth of child with congenital heart disease, depending on type of lesion in the first child. When two siblings have the disease, the risk for third affected child may increase to 20-30%. The incidence figures for infants born to mother, who have congenital heart disease are similar to those for sibling. Thus parents having affected child require counselling regarding the incidence of cardiac malformation in subsequent children.

The diagnosis of congenital heart disease can be made clinically, but electrocardiogram and skiagram of chest are necessary to support the clinical diagnosis. By the advent of echocardiography, various anatomical details can be made out without any invasive procedure.

Thus, by seeing the incidence of congenital heart disease in different parts of the world, it is obvious that this problem is quiet big and must be involving a significant number of children in Bundelkhand area. As no study has been made in this field, so far, in this area, present study was planned to work over the congenital heart disease in this area with following aims and objectives :

1. To find out the prevalence of congenital heart disease in Bundelkhand region.
2. To study the various types of congenital heart diseases and their incidences.
3. To find out the sex incidence of various congenital heart diseases.
4. To see association of congenital heart disease with other congenital defects.
5. To see the development delay in infants with congenital heart disease.
6. To see for the complications of disease in infants with congenital heart disease.
7. To see incidence of congenital heart patients probably affected by environmental factors, like diseases, medication or overexposure of radiation in mothers of affected children during gestation.

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REVIEW OF LITERATURE

REVIEW OF LITERATURE

INCIDENCE AND PREVALENCE :

An acceptable estimate of incidence of congenital heart disease in general population is not yet available. McKeown and Record (1960) & Carter (1961) considered that total incidence of all major malformations is 2.4% and those of heart form one quarter of these i.e. 0.6%. Malpas (1937) found only 10 patients with heart defects among 13,964 (0.7 per thousand) and Deporte and Parkhurst (1945) noted 142 in 300,795 births (0.5 per thousand). Sampson et al (1938); Rauh (1939) and Weiss (1941) gave estimates between 0.14 and 0.16 per thousand by examination of school children.

McMahan et al (1953) obtained the information about all cases of congenital heart disease born to Birmingham mothers in the years 1940-49. Diagnosis was confirmed at necropsy or by operation or by a consultant physician or by certification of cause of death. Incidence during whole period was 3.17 per thousand total births and 3.23 per thousand live births, with 1 per thousand still births.

Muir performed a necropsy study at Singapore in decades of 1948-57, found that congenital heart disease accounted for 2.2 percent of all necropsies, and for 8.4% of the necropsies done on children under the age of 10 years. The minimum mean incidence was shown to be atleast

1 per 1000 birth and for all still births, 1.7 per 1000. In America, the incidence was found to be 6.8 (Harris & Steinberg, 1954) and 6.5 (McSantosh et al, 1954) per thousand).

NERICP (New England Infant Cardiac Programme) figures shown incidence of congenital heart diseases to range between 1.5 per 1000 to 2.48 per 1000, average being 2.08/1000 live birth during 1969-1974 & 2.43/1000 live birth during 1975-1977.

Carlgren et al (1981) found it to be 9 per 1000 new born after studying all infants born in 1981 in Sweden by using different registries : Swedish Registry of Congenital Malformation, Medical birth Registry. Registry of Death Certificates and Child Cardiology registry.

In the India, Hadley et al (1958) in a series of 2000 necropsies at Vellore, South India found the incidence to be 1.3%.

Padmavati & Datey (1968), on studying prevalence of types of heart diseases in India found the distribution of congenital heart disease in Indian city hospitals to be 4.8% in Delhi (1951-55), 3.6% in Amritsar (1953), 6.3% in Bombay (1952-56), 2.3% in Madras (1946) and 1.6% in Lucknow (1953). This dealt with a very selected population group and was mostly from large hospitals attached to medical schools. They are however, more reliable because of more accurate diagnosis.

The incidence of different types of congenital heart disease, seems much the same as in other parts of world, found on various necropsy studies.

Dry et al (1948) found the incidence of various lesion as :-

VSD	-	12%
ASD	-	20%
PDA	-	15%
Coarctation aorta	-	12%
Fallot's tetralogy	-	9%
TGA	-	7%
Others	-	30% including truncus arteriosus great vessels anomaly, common AV canal etc.

McMohan et al (1953) found the following incidence in Birmingham (U.K.) :-

VSD	-	15.3%
ASD	-	8.6%
PDA	-	17.6%
Coarctation of aorta	-	10.6%
Fallot's tetralogy	-	4.0%
TGA	-	14.9%
Truncus arteriosus	-	6.3%
Others	-	22.5%

Gibson (1956) described distribution of different lesions as follows in London :-

VSD	-	8.5%
ASD	-	10.0%
PDA	-	4.5%
Fallot's tetralogy	-	20.5%
TGA	-	15.5%
Coaretation	-	4.0%
Pulmonary stenosis	-	2.0%
Aortic stenosis	-	2.5%
Fibroelastosis	-	7.5%
Others	-	25.5%

Muir (1959) gave the distribution pattern from a necropsy study of singapore to be -

VSD	-	23.1%
ASD	-	11.9%
PDA	-	11.4%
Coarctation	-	4.6%
Fallot's tetralogy	-	10.4%
TGA	-	5.1%
Aortic stenosis	-	2.4%
Pulmonary stenosis	-	8.0%
Others	-	22.8%

Keith et al (1978) reported the incidence of specific (major congenital defect in different age groups as following :

Defect		Percentage
VSD	-	28.0%
ASD	-	10.3%
Pulmonary stenosis	-	9.9%
PDA	-	9.8%
Fallot's tetralogy	-	9.7%
Coarctation of aorta	-	5.1%
TGA	-	4.9%

These comprises of 85% of congenital heart disease. Others are hypoplastic congenital heart syndrome. Total anomalous pulmonary venous drainage, Tricuspid atresia, Truncus arteriosus and other rare defects.

In India Vijay Priya et al (1979) conducted a clinical, hemodynamic and angiocardiographic study in 18 cases of cyanotic congenital heart disease. Fallot's tetralogy was found to be most common.

Kinare et al (1981) described the pattern of anomalies in 270 autopsied cases of congenital heart disease during first year of life in the department of Cardio-pathology, KEM hospital, Parel, Bombay during the year 1960 to 1979. Commonest during neonatal period was fetal coarctation (14%), followed by transposition complexes

(12.1%) and mitral and / or aortic atresia complexes (10.2%). Tetralogy of Fallot's (11.8%) was commonest during first year of life followed by transposition complexes (11.6%) and coarctation (7.7%). Associated non cardiac anomalies were present in 9.2% cases with a higher frequency of gastrointestinal anomalies.

In an other autopsy study done at PGI, Chandigarh from January, 1971 to December, 1974 by Banerjee et al in 250 infants of less than one mohth of age, 19 showed major cardiac anomalies. Clinical & necropsy records of them were reviewed and information regarding period of gestation at birth, age at death, sex, cause of death and associated pathology and extracardiac malformation was recorded. Hypoplastic left heart syndrome was the commonest and was seen in 6 (31.5%), Right sided obstruction in other 6 (31.5%), TGA in 2 and acyanotic heart disease in 5. Extracardiac malformation were present in 6 (31.5%) of which 4 were associated with hypoplastic left heart syndrome.

Campbell (1965) also found VSD to form 20% of total, then PDA, ASD, Coaretation of aorta, Pulmonary stenosis, Fallot's tetralogy & TGA are each responsible for about 10% and aortic stenosis for about 5%, this makes 85% of total, leaving 15% for other less common malformations.

Their distribution among older children shows some striking differences because of heavy early mortality especially in Ist year of life. VSD, ASD, PDA and PVS, each form about 15% of total, Fallot's tetralogy 12% and coarctation and aortic stenosis each 6% leaving 16% for other less common malformation. The incidence in general population of older children will be lower, about 3.6 per 1000. For ASD this is more than twice as high as the 0.2 per 1000 found by Seldon et al (1962) among the Australian population aged 15-65 yeers.

Scott et al (1984) reviewed the diagnosis and age at presentation of 1665 infants with symptomatic heart disease, who were admitted to Brompton hospital, London during the period 1973-82. The frequency of certain conditions had changed during the period of the study. Complete TGA and critical aortic stenosis had become less common where as frequency of right ventricular outflow tract obstruction, and of critical pulmonary stenosis had increased.

Most of the cyanotic infants presented during the first 14 days of life i.e. at the time of ductal closure. As expected this was also true of other duct dependent circulatory disorders as coarctation and interrupted aortic arch. Acyanotic infants with potentially large left to right shunts tended to present during the second month of life when the pulmonary vascular resistance fall. The

study also emphasise that most symptomatic infants with heart disease present during the first two months of life.

CLASSIFICATION :

Congenital heart disease are classified in different ways given by various authors. Nadas & Fyler presented the classification based on anatomical lesion or without being hindered by division of cyanotic or noncyanotic disease.

ANATOMICAL CLASSIFICATION :

I. Communication between systemic and pulmonary circuits with dominantly left to right shunts.

A. Interatrial communication -

1. Patent foramen ovale;
2. Secundum atrial defect;
3. Endocardial cushion defect.

B. Interventricular communication -

1. Simple VSD with or without pulmonary arterial hypertension;
2. Complicated VSD with -
   - Pulmonic stenosis
   - Aortic regurgitation
   - Artial defect
   - PDA
   - Coarctation of aorta

3. Single ventricle syndrome
   - Absent or extreme hypoplasia of RV
   - Absent or extreme hypoplasia of LV
   - Absent septum

4. Left ventricle - right artial shunt

C. Communication between great vessels -
1. PDA Simple
2. PDA complicated with other defects
3. Aortopulmonary fenestration
4. Truncus arteriosus
5. Ruptured aneurysm of sinus of valsalva
6. Coronary AV fistula

II. Valvular or vascular lesion with a right to left shunt or no shunt at all.

A. Coaretation of aorta
B. Vascular ring
C. Aortic stenosis
D. Aortic runoffs
E. Hypoplastic left heart syndrome
F. Mitral stenosis
G. Mitral regurgitation
H. Cor Triatriatum
I. Pulmonary vascular obstruction syndrome
   - Primary
   - Secondary

J. Pulmonary stenosis
   - with intact septum
   - with VSD

K. Pulmonary atresia

L. Pulmonary regurgitation

M. Underdeveloped right ventricle
   - Tricuspid atresia
   - Pulmonary valve atresia

N. Right ventricular dysplasia syndrome
   - Ebstein's anomaly
   - Uhl's disease

III. The transpositions -

A. Complete transposition of great arteries (D transposition)

B. Corrected transposition of great arteries (L transposition)

C. Double outlet right ventricle

D. Double outlet left ventricle

E. Other transpositions

IV. Venous anomalies -

A. Anomalous pulmonary venous drainage
   - Complete
   - Partial

B. Systemic venous drainage anomalies

V. Intrinsic dextrocardias -

A. Without heart disease -

- Situs invertus totalis with L loop
- Situs solitus with D loop

B. With heart disease -

- Situs inversus
- Situs solitus
- Asplenia with situs symmetricus

VI. Levocardias With or without atrial inversion.

CLINICAL CLASSIFICATION :

(a) Behrman & Aughan classification :

This depends upon presence or absence of cyanosis.

I. Congenital heart disease with cyanosis. (Dominant right to left shunt).

1. Tetralogy of Fallot
2. Pulmonary atresia with or without VSD
3. Tricuspid atresia
4. Double outlet right ventricle with PS
5. TGA (L and D)
6. Total anomalous pulmonary venous return
7. Ebstein's disease
8. Truncus arteriosus
9. Single ventricle
10. Eisenmenger syndrome
11. Hypoplastic left heart syndrome
12. Pulmonary AV fistula

II. Congenital heart disease with little or no cyanosis.

1. VSD
2. ASD
3. Partial anomalous pulmonary venous return
4. Endocardial cusion defect
5. PDA
6. Aortico pulmonary septal defect
7. Coronary artery fistula
8. Pulmonary stenosis alone or with left to right shunt
9. Ruptured sinus of valsalva
10. Double outlet right ventricle
11. Coaretation aorta
12. Mitral stenosis or insufficiency, Mitral valve prolapse
13. Anomalous origin of coronary artery.

(b) Morgan Classification :

Morgan (1978) classified them on the basis of presence and absence of cyanosis clinically, estimation of pulmonary blood flow in chest film and electrocardiographic findings. This give a relatively short list of possible diagnosis interfairly high degree of accuracy as given in table.

TABLE

Classification of Congenital heart disease

CYANOTIC

Decreased Pulmonary blood flow

RVH	LVH	BVH OR LVH
.PS .PS + VSD (Tetralogy) .Pulmonary vascular obstruction (Eisenmenger's) .Ebsteins	.Tricuspid atresia	.Truncus arteriosus with hypoplastic pulmonary artery .TGA + PS

Increased Pulmonary blood flow

RVH	LVH OR BVH
.TAPVC .TGA .Hypoplastic Left heart syndrome	.TGA + VSD .Single ventricle .Truncus arteriosus .Tricuspid Atresia + TGA

Acyanotic

Normal pulmonary blood flow

RVH	LVH
.Coaretation .Mitral stenosis	.Coarctation .Aortic stenosis .Mitral regurgitation .Endomyocardial disease

Increased pulmonary blood flow

RVH	LVH
.ASD .All left to right shunt with pulm. hypertension (PDA, VSD, ASD)	.PDA .VSD .Arteriovenous fistula

RVH - Right ventricular hypertrophy
LVH - Left ventricular hypertrophy
BVH - Both ventricular hypertrophy
VSD - Ventricular septal defect
ASD - Atrial septal defect
PDA - Patent ductus arteriosus
TAPVC- Total anomolous pulmonary venous circulation
TGA - Transposition of great vessels
PS - Pulmonary stenosis

Etiology :

The etiology of congenital heart disease remains obscure in most cases. A multifactorial inheritance hypothesis is gaining increasing acceptance. Single mutant gene syndromes probably represent less than 1 percentaand gross chromosomal abnormalities less than 5 percent of total. In most instances there is combination of genetic and environmental influences.

The various factors based on several studies, which are responsible for causation of the congenital heart disease will be discussed here.

Genetic factors :

Campbell & Polani, 1961 b, mentioned that in few families ASD is inherited through an autosomal dominant gene of low penetrance and there may be few similar families with congenital aortic stenosis. Dominant Mendelian inheritance is also found in familial cardiomyopathy (Bishop et al). Medial necrosis of the aorta is often seen with arachnodactyly (Marfan's syndrome), which is also inherited as mendelian autosomal dominant manner.

Situs invertus is a good example of recessive mendelian inheritance for malformation of the heart as it is more than 10 times the level in first cousins of

propositus than expected in general population of Britain. Fuhrmann (1958) found 40% of monozygotic pairs but only 25% of dizygotic pairs were concordant for some cardiac malformation, most often of the same type. Thus it suggest some genetic predisposition.

The high degree of concordance between malformation in propositus and sib is readily explained by genetic factors.

Miller & Smith (1979) described few families in which inheritance and recurrence risk appeared to be monogenic. This implies that there may be single genes that control specific events in cardiac embryogenesis such as conotruncal septation (CTS). Mutation of such genes could lead to abnormalities in cardiac morphogenesis, resulting in spectrum of cardiac defects grouped in CTS malformation complex. This recognition is important in rendering genetic counselling in cases with strong family history for CTS defects. A higher recurrence risk should be considered rather than the used polygenic recurrence risk of 3% that is usually found. Evidence for a genetic control of CTS arise from genetic and embryologic studies of similar defects in Keeshond dog model.

Corone et al (1982) done a statistical analysis of pairs of congenital heart lesions observed in families with atleast two affected members. The concordant lesions were found in roughly 50% of cases for first degree relatives. The analysis of discordant lesions showed excess in same pairs (Tetralogy, VSD and TGA) highlighting clusters of lesions and suggesting that these lesions, although dissimilar anatomically have some common genetic origin as they all are consequences of conotruncus septation anomalies. Deficit in some discordant pairs may be a consequence of interaction between effects of genes (epistasis effect). This leads us to think of heart not as a genetic whole but rather is being constituted of different specific genetically determined segments. This concepts agrees with the embryologic "segmental approach" suggested by Van Praagh and Van Praagh (1982).

Familial Incidence :

Incidence of congenital malformation of heart is raised in relatives of affected individuals. The evidence is supported by various studies by Abbott, 1927; Degramaci and Green, 1947; Campbel, 1949 and Lavy et al, 1950.

McKeown et al (1953) found the incidence of congenital heart disease among sibs born after the first propositus to be 18 per 1000 approx six times the incidence in general population of birth (3.2 per 1000). Incidence in first cousins of propositi was also found to be slightly higher (4.3 per 1000).

The incidence also increases with consanguinity of parents in few cases. This support for Cockayne's Suggestion (1938) that this condition may be inherited as a recessive.

Campbell (1965) found a strong tendency for the malformations found in the sibs to be concordant with those in the propositi in 56%, partially concordant in 22% and discordant in only 22%. The number of non-cardiac malformation were about 2.3% which is about the percent of major malformations present in general population.

Incidence of malformation of heart in parents was about 0.3% except A.S.D. malformation in the children of propositi were (4.4%) which is higher than would be expected by chance, especially for cardiac malformations. Consanguinity of parents of propositi was also studied by him & Lamy et al and incidence was found about 1.6%. P.V.S. (2.7%) and A.S.D. (3.1%) have highest index of consanguinity except for situs inversus (5.6%).

Dr. Ray Anderson in 1977 reviewed his experience in 109 sets of twins and triplets with a variety of cardiac defects. He revealed concordance rate for cardiac defects of 8.2% in monozygous twins and 2.2% in dizygous twins. Noonan (1978) also found similar results and on pooling Uchida etal's and his twin study concordance rate of 8.8% was found in monozygous and 3.7% in dizygous twins from total of 88 twin pairs. This supports the current concept that most cardiac congenital malformations result from multifactorial inheritance rather than a single genetic trait. In the both studies percentage of monozygous twins was disproportionately high. Instead of expected 30%, Anderson reported 58% and Noonan found 44% to be monozygous. In contrast to seperate twins, conjoined twins especially thoracopagus twins have a high incidence of cardiac defects that are frequently concordant. Thus he infered that mechanical factors can be very important in etiology of congenital heart disease as both monozygous and conjoined twins have the same genetic constitution but a very high incidence of congenital heart disease in conjoined twins contrast with the lower incidence in monozygous twins.

Parental Age and Birth order :

Campbell (1965) found no evidence that birth order alone has any general influence on the production of malformation of the heart. Lemy et al (1957) found that, if maternal age was held constant, birth rank was significantly higher in the congenital heart disease group than in control for the maternal age group 25-29 and 30-34. In their series only significant finding about birth order was linked with maternal age, and more sixth and later children with VSD were born to mothers aged 35 - 39.

Maternal age was rather more important. Only in Tetralogy of Fallot's and VSD did maternal age have a significant influence. After excluding children with mongolism more children with Fallot's tetralogy were born to mothers aged 40 to 45 years. In ASD group more children were of older mothers of 35 and over. MacMahon (1952) also thought septal defects were more common among the children of older mothers.

Penrose (1955) pointed out that the difference between means of paternal and maternal ages are in some way more useful measurement, since an undue increase of paternal age suggests the possibility of a "failure to copy genes correctly" because of larger number of cell

division in male germ line. The mean paternal age exceeded the maternal age by more than 2.3 years expected from the general population (Penrose, 1957) and ranged from 2.87 in coarctation and 3.03 in ASD to 3.48 in PVS and 3.70 in VSD. Lemy et al, (1957) found the fathers a little older and mother a little younger in their congenital heart disease group than in the controls.

Rothman and Fyler (1976) analysed the data of more than 2000 children born in New England, who were diagnosed with a congenital heart defect before the first birthday an enrolled in MBRICP. Subjects with diagnosis of Down's syndrome were excluded. Positive trends in risk with increasing birth order were present for pulmonic stenosis and transposition of great vessels and negative trends was seen for PDA. PDA and TGA displayed a pattern of risk with increasing maternal age. VSD had an erratic pattern of high risk for infants born of mother age 20-24 and low risk for older mothers, with mother less than 20 showing intermediate risk.

Mitchell et al reported a 70 percent increase in risk for all congenital heart disease when comparing subjects with maternal age 38 or more to subjects with maternal age 37 or less after excluding subjects with Down's syndrome. With regards to specific defects, Polani and Campbell (1958),

Kenna et al (1965) and Campbell (1965) all found risk to tetralogy of Fallot to be associated with greater maternal age but only Kenna et al (1965) found birth order also to be associated with tetralogy. They also reported positive associations of birth order with PDA and maternal age with Pulmonic stenosis.

ASSOCIATED MALFORMATIONS :

MacMahon et al (1962) observed other malformation in 101 (21%) of the 488 subjects while doing a study in Birmingham. The commonest association is with mongolism in 6% of cases. The incidence of other defects were much higher in children with congenital heart disease than in general population. In decreasing order of frequency they were alimentry tract anomalies, skeletal, genitourinary and nervous system anomalies, among which maximum are with septal defects.

In the series of NERICP from July, 1968 to June, 1977. 642 infants (28%) had new cardiac anomalies in addition to congenital heart disease among 2,381 infants. The percentage of extracardiac anomalies especially those graded "severe" i.e. having major effect on the life or well being of amenable to therapy, was highest among infants with endocardial cushion defects. This is largely the

result of association of mongolism within the group. After excluding the mongols the percentage falls to 18% with infants with endocardial cushion defects. Truncus arteriosus, Secondum ASD or PDA had a high percentage, while those with pulmonary stresis and intact septum, TGA or a aortic stenosis had only a small percentage of associated anomalies.

Syndromes were not common overall, but were found in 20% of infants listed as having severe extracardiac anomalies, most commonly with PDA, Skeletal anomalies were most common anomalies in almost all types, anomalies of respiratory and central nervous systems were most frequent among infants with PDA. Gastrointestinal anomalies were seen most often amont infants with PDA or endocardial cushion defects. Urinary anomalies were most frequent with PDA, Coaretation or truncus arteriosus and those in heterotaxia.

Campbell (1965) revealed that non-cardiac malformation occurs much more often than would be expected by chance. In his group the proportion varied from 5% in ASD to 13% in coaretation and PVS and averaged 9%. In the series of Lemy et al (1957), the figure was about 16% and varied between 7% to 26% in their different groups. Wood (1956) says that other malformation are present in form 10% to 20%, the figure in clinical series higher one in those found at necropsy.

Greenwood et al (1975) seen that extracardiac anomalies occured in 25% of infants during first year of life. Often they are multiple and one third of affected infants have some established syndroms. The presence of extracardiac anomaly significantly increases the mortality in infants with congenital heart diseases.

When a patient has one malformation of heart he is more liable than others to have a second one. The proportion varied from 8% with coaretation to 15% of those with ASD and 21% with PVS. The average figure for all acyanotic groups was 13%, about 20 times the expected figures. This includes some well known associations as aortic stenosis or PDA with coaretation but excludes the two malformation that must be present in more cyanotic malformation as VSD and PVS in Fallot's tetralogy.

Among chromosomal anomalies trisomy - 21 i.e. mongolism is so often accompanied by malformation of heart especially septal defects, that these too might be caused, in part atleast, by the trisomy. Recently other syndromes with multiple malformation including most often VSD have been described. These were E_1 or chromosome 17 - 18 and D_1 chromosome 13 - 14 trisomy where VSD was present in 20 out of 22 cases of former and 9 out

of 15 cases of the latter syndrome (Taylor and Polani, 1964; Campbell and Goldwin, 1965). The association of turner's syndrome with coarctation of the aorta (Campbell and Polani, 1961), suggest that it too may be due to abnormal XO chromosome complement.

Cullum et al (1965) in California went through the death records between 1957 to 64 and found that 4.8 percent of all deaths due to congenital heart diseases were associated with Down's syndrome. The type of defects were VSD (32%) AV canal defect (24.5%), ASD (20.1%), PDA (11.5%), Tetralogy (10.8%) and others (8.6%).

Laursen (1975) investigated total 1504 children with congenital heart disease under age of 15 years and found 80 patient with Down's syndrome i.e. 5.1%. Among those VSD was most commonly found 49%). He also noted that eisenmenger's syndrome appeared at earlier age in mongeloid children with VSD compared to other ones.

Corno et al (1982) reported the unusual association of stenotic dysplastic pulmonary valve, peripheral pulmonary stenosis, right aortic arch, aberrent left subclavian artery and complex aortic coarctation in a boy with Noonan's syndrome. Congenital heart disease occur in 50% of patients with Noonan's syndrome but most frequent is dysplastic pulmonary valve.

Sex incidence :

The overall sex ratio between male/female is 55/45 - Carlgren (1959), 53/47 Mcmahon et al, 51/49 Gardiner and Keith (1951) and 52/48 Muir (1959). In NERICP series also male babies predominated at 53.7%, but decreased to 51.8% of the one year survivors. There was female infants (37.8%), which is not surprising since male infants tended to have more lethal cardiac lesions. The relative incidence of the high risk factors were 53.8% in females. In general mortality among male infants was about 5% greater than among female infants. Banerjee et al (1975) found it to be 1.7:1 in study done in PGI, Chandigarh.

The different distribution of the two sexes in so many common malformation of heart has been recognized for a long time. PDA is the only condition more common in the female sex at birth and through life. Pulmonary stenosis shows an equal incidence in both sexes and transposition (70%) and Fallot's tetralogy (60%) show excess of male cases (Campbell, 1965).

Brotmacher & Campbell,(1958) concluded that boys reversed the left to right shunt at an earlier age than girls and became cyanotic and died earlier; and suggested that this might be because boys are less willing to limit

their exertion to an appropriate standard. Campbell & Polani (1961 b) found that the sex distribution was about equal in first decade, but in the third decade and after the male/female ratio became 1 : 2 and remained at that level for ASD.

Sex incidence at birth & later

Condition	Percentage of male subjects		
	At birth (MacMahon et al & Carlgren)	Children 0-15 (Keith et al)	Children mostly over 5 & adults (Campbell)
ADA	40	31	27
ASD	50	40	34
Coaretation	50	65	62
Aortic stenosis	55	60	70
Pulmonary stenosis	55	-	50
VSD	59	50	30
Fallot's tetralogy	61	60	59
Transposition	73	68	-

ENVIRONMENTAL FACTORS :

Environmental influences during pregnancy may be implicated as the cause of congenital heart disease. Various infections, diseases, medication or over exposure of radiation in mother of propositi may be found. Rubella affecting the mother during the first trimester of pregnancy is most clearly proved environmental cause.

Julia Bell (1959) studied 421 cases of maternal rubella in early pregnancy. Various malformations as deafness, cataract, congenital heart disease, mental retardation, microcephaly and others can occur, either alone or in combination. Among the heart malformation, PDA was commonest (58%) than VSD (18%), PDA & VSD both in (6%), ASD, Tetralogy of Fallot's and pulmonary valve stenosis each occur alone in 6% of cases.

Campbell (1961) found rubella in first trimester of pregnancy to be best established of all causes, but it does not explains a large proportion of cases. The risk of abortion or of malformation is high, between 40% and 60% during the first four weeks, between 30% and 50% during the second four weeks, between 20% and 40% during third four weeks, still perhaps a little during fourth four weeks, but not increased late, than this. Maternal rubella may be responsible for between 1% and 2% of all malformation of the heart.

Other viral infections responsible for malformation following maternal infection during pregnancy are measles, chickenpox, whooping cough, herpes zoster and infectious hepatitis-only one or two instances of each was reported. Mumps, poliomyelitis, influenza and toxoplasmosis (Fraser, 1959) has been incriminated in isolated cases.

At the congress of congenital heart disease in London in July, 1960, population of all those cases caused by maternal rubella and other viral infections was thought to be less than 10%.

Michalls and Mellin, 1960, have also proved rubella virus to be teratogenous for heart. Teratogenecity of Coxsackie - B virus causing in later months of pregnancy was also proved by Kibrick et al, 1956, 1958. It can also give rise to congenital cardiopathies when infection occur during first month of gestation (Brown, 1966, 1969).

Alzamora et al, 1953; Chavez et al, 1953; and Epaime-Vela, 1967 proved that there is a higher incidence of congenital heart disease with Arteriovenous shunt in children born in regions at an altitude of above 3000 meters and over above sea-level, as compared with populations at see level comparing the cities of Junin and Lima in Peru and at altitude of Mexican Plateau. These suggest that hypoxia is a possible causal factor and this in turn is supported by experimental work of Ingalls et al (1952), who proved its teratogenic effect in mice by producing VSD. The frequent occurrence of congenital heart disease has been seen in children born of mothers with congenital heart disease. This could be due to an intrinsic factor related to sex or to an extrinsic factor represented by hypoxia of maternal tissues.

Several malformations of heart occured after pregnancies in which there had been severe bleeding or threatened abortion or in which injection of Corpus luteum or other preparations had been given to avoid miscarriage. Lamy et al (1957) also found a history of such episodes twice as often as in their control series.

Harlap et al (1975) interviewed pregnant women and mothers in West Jerusalem in 1966-68 and found 3.8% babies to be born after definite or probable administration of estrogen or pregesterones. 47 out of these 432 babies had one or more major or minor malformation - a rate of 108.8 per 1000, compared with 77.6 per 1000 babies with no history of exposure to hormones. There was excess of heart defects and other defects of blood vessel development in babies born to mother who probably took hormones in early pregnancy. An excess of malformation would be expected among babies born to mothers with threatened or previous miscarriage, whether or not they were also given hormones. ASD was found usually after administration of stilbesterol specifically. The defects reported approx. twice the expected among infants with prenatal exposure to hormones.

Rothman et al, 1979 took the history of oral contraceptive use, hormonal pregnancy tests, prescribed hormones and other drugs before or during pregnancy in Massachusetts, Boston and positive history was obtained from 390 mothers of infants with congenital heart disease and 1254 mothers of normal infants. A small positive association between hormonal exposure and cardiac malformation was found. The prevalence ratio of exposed to non-exposed being 1.5. No association was evident between hormones and trunco-conal or any other class of defect among the cases.

Nora et al, 1976 also reported association between maternal exposure to progesterone/oestrogen at the vulnerable period of embryogenesis and congenital malformation involving several systems including cardiovascular, skeletal, gastrointestinal and genitourinary. CVS anomalies were VSD in 5 (in 4 cases) with early spontaneous closure) PDA in 2 and TAPV in 1 case among 100 live born infant. The overall risk after hormone exposure seems to be 2-4 times that of general population.

Heinonen et al, 1977 showed positive association between cardiovascular defects and phenobarbitone, phenothiazines and hormones as well.

Aminopterine (aborificient), busulphan and thalidomide all have produced malformations in man as well as in animals; but no drug has so far been incriminated as a cause of many malformation of the heart (Campbell, 1965).

Nora & Nora in 1977 reported that maternal rubella, ingestion of Thalidomide, folic acid antagonist during early gestation and chronic alcohol abuse are various environmental factors known to interfere with normal cardiogenesis. Maternal therapy with anti-convulsant agents especially Diphenylhydantoin and Trimethadione, Dextroamphetamine, Lithium chloride, Progesterone/Oestrogen, Warfarin and overexposure to radiation are associated with high incidence of congenital heart disease. Maternal lupus erythematosus during pregnancy has been linked to congenital complete heart block.

Cox (1964) found that malformation in general were twice as common in children of mothers who had been exposed to frequent X-ray examinations for congenital dislocation of hip.

Deprivation of vitamins and other abnormality in diets, can produce malformations of many types. Wilson et al (1953) found the malformation of aortic arches and bulbus and defects of ventricular septum

occured if mother rats were kept on diets difficient in vitamin A. The poor diet and malnutrition of many mothers have led to such malformations. Experimental study of extrinsic factors has proved the following other agents to cause congenital cardiopathies - Hypervitaminic 'A' diet (Kalter and Warkany, 1961), deficiency of ptereilglutamin acid (Nelson, 1960; Baird et al, 1954), deficiency of riboflavine (Kelter and Ulerkany, 1957; Nelson et al, 1956), Tryptan blue (Fox and Goss, 1958; Wilson, 1955).

Seasonal influences :

The season of conception provides some environmental factors that may have an influence on causation of malformations but it is easier to talk about the season of birth. Record and Mckeown (1953) in Birmingham, found a seasonal increase of PDA for girls, but not for boys from May to December, with a peak in July and August with coarctation of aorta, more boys were born in March and April and fewer in September and October. With PVS three times as many boys were born in July - September quarter as in April - June quarter, but girls were evenly distributed. With ASD nearly twice as many boys were born in January - March quarter then other but girls were evenly distributed.

Symptomatology :

Certain congenital anomalies are of no clinical importance since they produce neither subjective symptoms, physical signs nor other objective abnormalities. These include bifid apex of heart, persistant left superior vena cava and most instances of patent foramen ovale and of anomalous septa and chordae. Other anomalies produce neither symptoms, nor signs but are the site of serious complications eg. bicuspid aortic valve is often complicated by calcific aortic stenosis and by bacterial endocarditis. In pure dextrocardia with situs invertus there are no symptoms but physical signs and roentgenoscopic and electrocardiographic examinations reveal a characteristic picture. There may be abnormalities in blood pressure as in cases of coarctation of aorta or PDA which may be associated with altered circulatory dynamics as those in cases of free aortic regurgitation.

Dyspnea or exertion is a common symptom and often appears early in course of the disease. A striking form of dyspnea is that which appears in paroxysms and is usually associated with intensification of cyanosis (Anoxic Spell). In many cases exertional dyspnea is secondary to pulmonary congestion associated with left sided heart failure. Often it is due to excessive arterial oxygen unsaturation

especially following exercise with consequent anoxia of carotid sinus and respiratory centre. In cases of cyanotic congenital heart disease, there is in addition a further intensification of arterial hypoxemia, a rise in arterial PCO_2 and a fall in pH.

Squatting especially after exercise is common in certain form of congenital heart disease with cyanosis and characteristically with tetralogy.

Cough complicates dyspnea when later is due to pulmonary congestion but both may be due to tracheal compression by a double aortic arch, anomolous vessel or aberrent vessel or congestion of abdominal viscera from right sided heart failure. Dysphagia and hoarseness may occur due to compression of Oesophagus and recurrent laryngeal nerve respectively.

Poor feeding, subnormal gain in weight, constipation, excessive fatigability and weakness are relatively common.

Cerebral symptoms including faintness, dizziness and headache are observed occasionally and rarely syncope, convulsion, delirium and coma especially in patients with cyanosis. These are due partly to cerebral hypoxia especially during exertion and partly to polycythemia with consequent circulatory stasis or complicating cerebral thrombosis or hemorrhage. These may also be due to

inadequate cerebral blood flow in case of aortic stenosis or to ruptured congenital aneurysm of circle of willis in coarctation of aorta or to cerebral abscess in cases of septal defects and paradoxical embolism. Stokes adams attacks can occur in cases of congenital heart block.

Cardiac pain may be related to inadequate coronary perfusion during exercise in aortic or severe pulmonary stenosis.

Vascular disturbances in extremities including coldness, numbness and tingling or pain may appear in patients with or without cyanosis or polycythemia. These may occur in coarctation of aorta. Malnutrition and under development may result from deficient peripheral blood supply.

Sudden death is most likely to occur in cases of idiopathic myocardiopathy, subaortic stenosis or ebstein's disease. Otherwise death in congenital heart disease is due to congestive heart failure, anoxia, cerebral complications, vascular thrombosis, bacterial endocarditis or some intercurrent illness.

Cyanosis and clubbed finger are also found, but it is erroneous to believe that these are found in majority of cases. Abbott, in series of 1000 cases found cyanosis of varying intensity in 257 and in 124 it appeared terminally. Clubbing was found in only 132.

A secondary polycyathemia occurs frequently in cases with cyanosis. This occurs to compensate for oxygen unsaturation of blood. R.B.C. count usually varies between 6 and 7 milions / cm. mm. Haemoglobin is correspondingly elevated to 110 to 130 percent with moderate and may be upto 200% with extreme polycyathemia. The hematocrit usually exceeds 53 percent and often exceeds 65 percent.

Physical and mental development :

Linde et al. 1971 studied 319 children with approximately equal sex and compared intellectual development of cyanotic and acyanotic children with acnagenital heart disease to that of both their normal sibling and randomly selected well babies. Cyanotic patients scored lower at all ages particularly in earlier years with tests involving gross motor abilities in Gesell and Cattell developmental examinations. Correlation of low test scores to physical incapacity tended to disappear at later ages when stanford - Binet test were given. They also noted that delay in use of first words and first phrases was much less marked in children with heart disease. Cyanotic children were significantly more incapacitated than acyanotic ones. Early motor performance deficit in handicapped child may cause underestimation of intellectual potential.

Ruth et al (1982) also assessed the mental and motor development of 173 infants with congenital heart disease by assessment of Bayley Scale of infants development and clinical neurological examination. Developmental delay i.e. Developmental Index Score of below 80 was manifested by 25% of all infants. About half of them had delay on mental scale alone and half on both mental as well as motor scales. The presence of congestive heart failure was found to be significantly associated with both mental and motor developmental delay. Hypoxemia and hospitalisation were associated with delayed motor development.

Life expectation of children with congenital heart disease :

Macmahon et al (1952), estimated the life expectation at birth based on all cases of congenital heart disease (633) identified at birth or later in population of 199,418 total births. They suggest that of 10 affected children born alive, 2 die by the end of first week, 3-4 by the end of first month and 6 by the end of first year. Between 3 - 4 survive upto ten years.

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MATERIAL AND METHODS

MATERIAL AND METHODS

Study population comprised of the patients of various types of congenital heart diseases attending the Out Patient Department of Pediatrics or admitted in Pediatric ward of Maharani Laxmi Bai Medical College Hospital, Jhansi during September, 1989 to August, 1990. Detailed history and clinical examination was conducted and various investigations were done to confirm the diagnosis.

In this study we used the same definition of congenital heart disease as Mitchell (1957), who defined it as "a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance". Abnormalities of systemic veins and systemic artery branches were excluded but patent ductus arteriosus was included.

In the study, the classification of congenital heart disease given by Beverly, C. Morgan (1978) was used. The lesion were first classified on the basis of presence of or absence of cyanosis clinically. Later according to the pulmonary vasculature in chest roentgenogram, they were further divided into lesion with decreased or increased or normal pulmonary blood flow, and then according to electrocardiographic findings.

Where more than one lesions were associated, they were classified according to dominant lesion.

Detailed interrogation was made pertaining to presenting complaints, history of present illness and history of past illness. A detailed history regarding any maternal illness, history of taking medication or exposure to radiation during gestation was asked. History regarding any abortion or still birth or any other sibling or family member suffering from any congenital heart disease, consanguinity, elderly or teenage mother, and birth order were also taken.

Developmental history was recorded in all the spheres. Gross-motor, fine motor, social and speech mile stones attained till date was recorded in every case. In four developmental fields, quotient was calculated by dividing the developmental age from chronological age and them multiplying this value by 100 (Prabhakar and Kumar, 1983).

Every patient was examined in detail, including general examination, anthropometric measurements, detailed cardiovascular examination and other systems to look for other congenital defects.

Particular emphasis was paid during cardiovascular examination. This included examination of radial as well as other pulses, blood pressure and examination of precordium. Careful inspection, palpation, percussion and auscultation was done in each case to see for site of appex beat, any thrill, other pulsations, heart sounds and various murmurs.

While investigating the case routine haemotological studies were done in each case. At the same time chest skiagram P.A. view, and if needed lateral and oblique & barium swallow studies were also done. Electrocardiogram was taken in all the cases. We also included echocardiographic findings of the patients, who already had it with them while they attended the hospital or shown to us in follow up after getting it done at hospitals equipped with such facility. We had forty such cases.

Diagnosis was made and later defects were classified under various groups as shown in table.

TABLE

Classification of Congenital heart disease

CYANOTIC

Decreased Pulmonary blood flow

RVH	LVH	BVH OR LVH
.PS	.Tricuspid atresia	.Truncus arteriosus with hypoplastic pulmonary artery
.PS + VSD (Tetralogy)		.TGA + PS
.Pulmonary vascular obstruction (Eisenmenger's)		
.Ebsteins		

Increased Pulmonary blood flow

RVH	LVH OR BVH
.TAPVC	.TGA + VSD
.TGA	.Single ventricle
.Hypoplastic Left heart syndrome	.Truncus arteriosus
	.Tricuspid Atresia + TGA

Acyanotic

Normal pulmonary blood flow

RVH	LVH
.Coarctation	.Coarctation
.Mitral stenosis	.Aortic stenosis
	.Mitral regurgitation
	.Endomyocardial disease

Increased pulmonary blood flow

RVH	LVH
.ASD	.PDA
.All left to right shunt with pulm hypertension (PDA, VSD, ASD)	.VSD
	.Arteriovenous fistula

RVH - Right ventricular hypertrophy
LVH - Left ventricular hypertrophy
BVH - Both ventricular hypertrophy
VSD - Ventricular septal defect
ASD - Atrial septal defect
PDA - Patent ductus arteriosus
TAPVC - Total anomalous pulmonary venous circulation
TGA - Transposition of great vessels
PS - Pulmonary stenosis

Patients having VSD were again classified in mild, moderate or severe type according to clinical assessment of the defect. The patients with mild or small VSD were those, who presented later in infancy with no or mild symptoms, normal split second heart sound and without any flow murmers. E.C.G. and X-ray were normal. Patients with moderate type of defects had moderate symptoms with normal or narrow split of second heart sound, with or without flow murmer and mild abnormalities of X-ray and ECG with or without pulmonary hypertension. In severe type, patients presented early in infancy usually with congestive cardiac failure. The flow murmers, narrow split and pulmonary hypertension were usually present.

In X-ray there was significant cardiomegaly, prominent pulmonary artery and increased pulmonary vasculature. It was possible to confirm the size by echocardiography in seventeen of the cases out of 30 cases.

At last all the findings were tabulated and analysed statistically.

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OBSERVATION

OBSERVATIONS

The present study entitled 'Study of congenital heart diseases in Bundelkhand region' was performed in Maharani Laxami Bai Medical College Hospital, Jhansi from September, 1989 to August, 1990. Total number of children, who attended the pediatric out patient's department were 12,810 and out of them total 186 children were found to be having congenital heart disease. Out of these, 62 children could be investigated or followed up, thus comprised the study population. Thus prevalence of congenital heart diseases in pediatric diseases on the basis of available hospital records,was 1.45%.

The age distribution of the patients under study is shown in table 1. The youngest one was of 7 days, while the oldest was of 13 years. 29 patients (46.8%) were upto one year of age and among them 4 patients (6.5%) were of less than one month of age, 19 patients (30.5%) were of one to six months age group and 6 patients (9.7%) were of six months to one year age group. 'One to two years' age group was having 6 patients (9.7%) and 'two to three years' age group was having 5 patients (8.1%). Both 'three to four years' and'four to five years'age groups were comprised of 3 patients (4.8%) each. Patients between 'five to ten years' were 10 (16.2%) while those of more than ten years of age were 6 (9.7%).

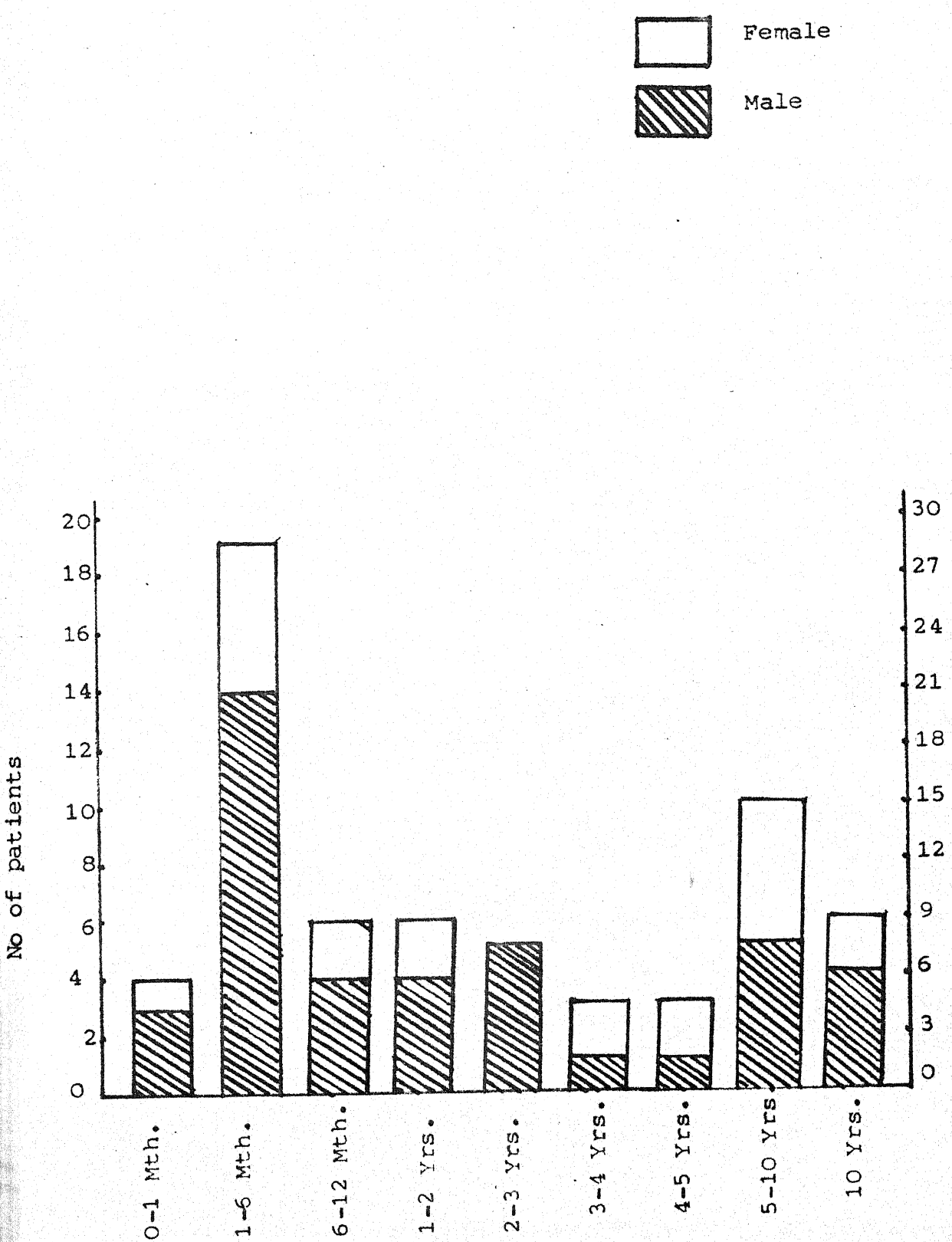


Figure - 1 AGE AND SEX DISTRIBUTION

Table - I

Age and Sex distribution

Age of presentation	Number of patients			
	Male	Female	Total	Percentage
0 - 1 month	3	1	4	6.5
1 - 6 month	14	5	19	30.5
6 - 12 month	4	2	6	9.7
1 - 2 year	4	2	6	9.7
2 - 3 year	5	0	5	8.1
3 - 4 year	1	2	3	4.8
4 - 5 year	1	2	3	4.8
5 - 10 year	5	5	10	16.2
7 10 year	4	2	6	9.7
Total	41	21	62	100.0

Out of all the sixty two patients studied, 41 (66%) were male and 21 (34%) were female. Thus male female ratio of present study was 1.94 : 1. The over all sex distribution in different age groups is also shown in table - I.

Eventful antenatal history was recorded in 9 (14.4%) out of 62 patients of congenital heart disease. Among them history of taking oestrogen/progesterone preparations in fourth or fifth month of antenatal period was present in 4 (6.5%), history of taking medication during gestation (anti-tubercular drugs) in 2 (3.2%), history of radiation

during fourth month of antenatal period in 1 (1.6%) and history of toxemia of pregnancy was present in 2 cases (3.2%). Distribution of all these patients are shown in table II.

Table - II

Patients with eventful antenatal history

Antenatal events	Number of patients	Percentage
Oestrogen/Progesterone intake	4	6.5
Drug intake	2	3.2
Exposure to radiation	1	1.6
Toxemia of pregnancy	2	3.2
Total	9	14.5

Among the all sixty two patients the family history of congenital heart disease was present in 3 cases (4.8%). One of them had same lesion as that of the affected patient i.e. VSD. In two other cases, there is history suggestive of some heart disease in two other siblings, who expired during first month by becoming blue.

History of prematurity was found in 3 cases (4.8%) among all sixty two patients. All the three were born of about twenty eight to thirty weeks of gestation and had VSD.

Associated congenital anomalies were found in 4 (6.4%) cases out of sixty two. Features of mongolism was found in 2 cases (3.2%). Both of them were associated with VSD. One other child of ASD was having imperforate anus (high type) and was having colostomy. In one case of VSD, hydrocephalus and cystic hygroma were found to be present.

Modes of presentation in the cases is shown in table III.

Table - III

Modes of presentation

Modes of presentation	Number of patients	Percentage
1. Failure to thrive	16	25.8
2. Recurrent chest infections	20	32.3
3. Cough & breathlessness	30	48.3
4. Exertional dyspnoea	15	24.2
5. Feeding difficulty	16	25.6
6. Blue coloration of lips	5	8.1
7. History of cyanotic spells	5	8.1
8. Squatting	4	6.4
9. Palpitations	2	3.2
10. Oedema feet	2	3.2

The most common symptoms were of cough and breathlessness, which were found in 30 (48.3%) out of sixty two. History of attacks of recurrent chest infections was found in 20 (32.3%) patients. Failure to thrive was the complaint of 16 (25.8%) patients. Dyspnoea on exertion was found in 15 (24.2%) patients. In 16 (25.6%) of them feeding difficulty was found. 5 patients out of 12 of cyanotic group (41.6%) complained of blue discolouration of lips and nails which became more marked on crying, rest were found to have cyanosis on examination.

History of cyanotic spells was present in 5 (41.6%) patients out of 12 cyanotics. History of assuming a characteristic posture after exertion by patients i.e. squatting was found in 4 (33.3%) cases of all cyanotics and all of them were having tetralogy of Fallot. In two cases of older age group, complaint of palpitations and in another two, complaint of oedema feet was present. Clubbing was found in 7 out of 12 cyanotics & one of acyanotic group. Congestive heart failure was found in 19 (30.4%) patients out of all. 40% of cases of VSD presented with congestive cardiac failure and other cases of CHF were of TGA (2), coarctation of aorta (1), Ebstein anomaly (1), Pulmonary stenosis (1) and ASD (1). All the children of cyanotic type had anaemia and the PCV ranged from 35% to 50%.

The distribution of patients according to clinical types of congenital heart disease is shown in table IV.

Table - IV

Differential distribution of congenital heart disease

Type of malformation		No.of cases	Percentage
A. Acyanotic	1. Ventricular septal defect	30	48.2
	2. Atrial septal defect	8	12.9
	3. Patent ductus arteriosus	5	8.1
	4. Endocardial cushion defect	1	1.6
	5. Coarctation of aorta	1	1.6
	6. Bicuspid aortic valve with stenosis	1	1.6
	7. Pulmonic stenosis	1	1.6
	8. Aortic and mitral insufficiency	1	1.6
	9. Dextrocardia	3	4.8
Total		50	80.6
B. Cyanotic	1. Tetralogy of fallot	5	8.1
	2. Transposition of great arteries	3	4.9
	3. Ebsteins anomaly	1	1.6
	4. Tricuspid atresia	1	1.6
	5. Hypoplastic left heart syndrome	1	1.6
	6. Anomalous systemic verious drainage	1	1.6
		12	19.4

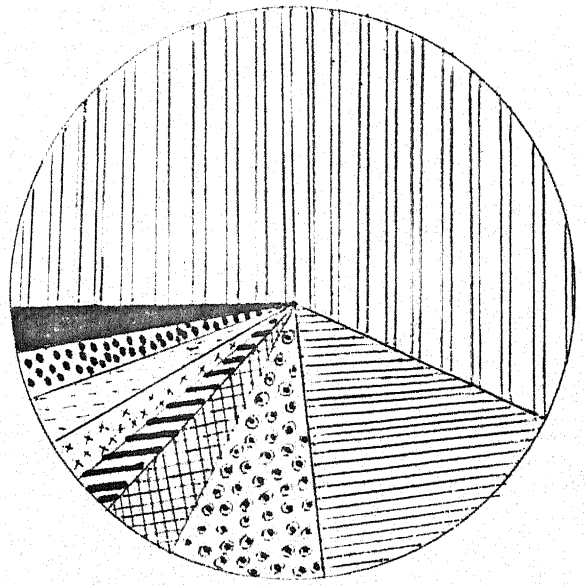


Figure - 2 DISTRIBUTION OF ACYANOTIC PATIENT

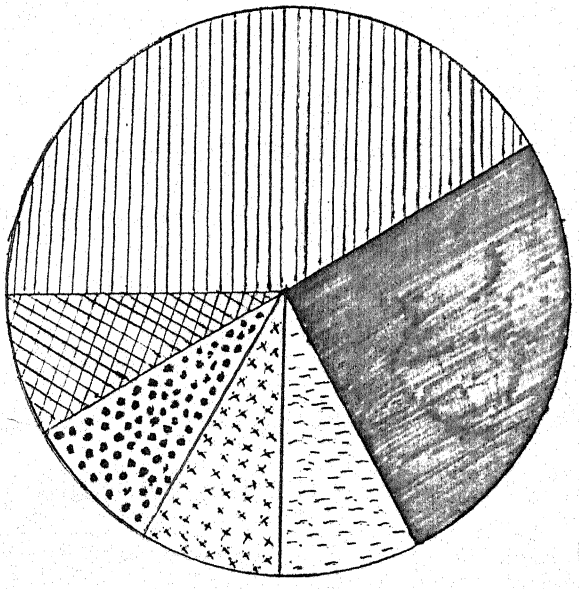


Figure-3 DISTRIBUTION OF CYANOTIC PATIENTS

The patients belonging to acyanotic group were 50 (80.6%) out of all 62 patients. In cyanotic group, there were only 12 (19.3%). Among all acyanotic patients VSD was most common. It was seen in 30 (48.2%) patients. The next common distribution in acyanotic patients was of ASD, which was found in 8 (12.9%) patients. Patent ductus arteriosus was next in sequence having 5 (8.1%) patients. Other abnormalities i.e. endocardial cushion defect, coaretation of aorta, bicuspid aortic valve, pulmonic stenosis, aortic and mitral insufficiency had one (1.6%) patient in each group.

Among the 12 cyanotic cases, tetralogy of fallot was most common, present in 5 (8.1%) patients. In 3 (4.8%) patients transposition of great arteries was found. Ebstein anomaly, tricuspid atresia, hypoplastic left heart syndrome and anomalous systemic venous drainage were less common abnormalities. Only one patient (1.6%) of each type was found during our study in echocardiography.

The sex distribution was found different in different type of lesions. Its distribution among acyanotic patients is shown in table V.

Table - V

Sex distribution among acyanotic patients

Type of malformation	Number of cases		
	Total	Male	Female
1. V.S.D.	30	21	9
2. A.S.D.	8	4	4
3. P.D.A.	5	2	3
4. Endocardial cushion defect	1	-	1
5. Coaretation of aorta	1	1	-
6. Bicuspid aortic valve with stenosis	1	-	1
7. Pulmonic stenosis	1	1	-
8. Aortic and mitral insufficiency	1	1	-
9. Dextrocardia	2	1	1
Total	50	32	18

All the conditions were found to be more common in males except for PDA, endocardial cushion defect and aortic stenosis in acyanotic group of patients. In VSD among all 30 patients 21 were male and in ASD among all 8 patients 4 were male, while in PDA among 5, two were male and 3 were female. In other groups no typical sex predominence could be commented as there were very few patients in each group.

The sex distribution among cyanotic patients is shown in table VI. In tetralogy of fallot, there was definite male preponderance as all 5 patients recorded were male. Transposition of great arteries was found to be common in males as there were 2 affected males and one female. In other types only one patient was found of each group and all of them were male except for hypoplastic left heart syndrome.

Table - VI

Sex distribution among cyanotic patients

Type of malformation	Number of cases		
	Total	Male	Female
1. Tetralogy of fallot	5	5	-
2. Transposition of great arteries	3	2	1
3. Ebstein's anomaly	1	1	-
4. Tricuspid atresia	1	1	-
5. Hypoplastic left heart syndrome	1	-	1
6. Anomalous systemic venous drainage	1	1	-

Ventricular septal defect was present in large number of cases. This was also categorised into mild, moderate and large sized VSD. The distribution is shown in table VII. The mild VSD was suspected in 8 cases (26.7%), moderate in 7 (23.3%) cases and large in 15 (50%) cases out of all 30 affected children.

Table - VII

Distribution of VSD according to clinical size and septal defect

Type of VSD	No. of cases	Percentage
Mild	8	26.7
Moderate	7	23.3
Large	15	50.0

The mean developmental quotient was calculated in all cases and analysed statistically by student 't' test both in motor and mental fields.

Mean developmental quotient in congenital heart disease is shown in table VIII.

Table - VIII

Mean development quotient in congenital heart disease

Type of heart disease	No.of patients	Mean Developmental Quotient	
		Motor	Mental
1. Acyanotic heart disease	30	78*	86.4 **
2. Cyanotic heart disease	12	66*	80.4 **
't'		2.01*	.225**
P		$< .05$	7 .8
d.f = 60			

The least retardation of development was seen in manipulative and social mile stone fields, especially in acyanotic type. Developmental quotient was 78 in acyanotic and 66 in cyanotic group of patients in motor fields. The difference was statistically significant ($p < .05\%$) while in social and speech, it was (86.4%) in acyanotic and (88.4%) in cyanotic group the difference was not significant ($p > .8\%$).

All the patients were classified into various grades of malnutrition by their weight as classified by Indian academy of Pediatrics. There classification is shown in table - IX.

Table - IX

Malnutrition in congenital heart disease

Degree of Malnutrition	Total		Acyanotic	Cyanotic
	No. of cases	%		
Grade - I	17	27.0	16	1
Grade - II	13	21.0	9	4
Grade - III	4	6.5	3	1
Grade - IV	4	6.5	4	0
Normal	24	39.0	18	6

39% of patients had normal weight for age while 27% fall in I degree, 21% in II degree, 6.5% in III degree and 6.5% in IV degree malnutrition. Only 13% i.e. 8 patients were of severe malnutrition.

The correlation of clinical and electrocardiographic finding in acyanotic heart disease patients is shown in table - X. Among 30 cases of VSD, in 14 cases ECG was normal. RVH was found in 9, in which 3 cases were also having associated pulmonary stenosis and 2 were having pulmonary hypertension. BVH was found in 4 cases in which one was also having pulmonary stenosis and an other, pulmonary hypertension LVH was present in 2 cases of simple VSD.

In total 8 cases of ASD, RVH was present in six cases and normal ECG was present in a single case. In 3 cases both ASD and VSD were visible in Echo. Two of them had RVH and one had BVH.

There were total 5 cases of PDA, one case also had pulmonary hypertension and in two, VSD were also found to be present in Echo. LVH was present in 3 and BVH in 2 cases.

There was a single case of endocardial cushion defect of common atrioventricular canal type as seen an Echo, which was having left axis deviation. In cases of coarctation of aorta and aortic and mitral insufficiency, LVH was found. In a case of aortic stenosis, bicuspid aortic valve and VSD was also seen on echo. She was having BHV on ECG. In a single case of pulmonary stenosis, RVH was there.

Table - X

Correlation of clinical and ECG findings in patients

Clinical diagnosis	Number of cases				
	Total no.	With RVH	With LVH	With BVH	Normal
1. VSD -					
a) Simple VSD (30 cases)	23	4	2	4	13
b) VSD with PS	4	3	-	1	-
c) VSD with pulmonary hypertension	3	2	-	1	-
2. ASD -					
a) Simple ASD (6 cases)	4	3	-	-	1
b) ASD with PS	1	1	-	-	-
c) ASD + VSD	3	2	-	1	-
3. PDA -					
a) Simple PDA (5 cases)	2	-	1	1	-
b) PDA + PH	1	-	1	-	-
c) PDA + VSD	2	-	1	1	-
4. Endocardial cushion defect	1	-	-	-	1
5. Coarctation of aorta	1	-	1	-	-
6. Aortic stenosis	1	-	-	1	-
7. Aortic and mitral insufficiency	1	-	1	-	-
8. Pulmonic stenosis	1	1	-	-	-

The clinical and ECG findings of cyanotic patients are correlated in table XI. There were 3 Echocardiography proved cases of TGA. Two of them had RVH and one was having LVH which also had pulmonic stenosis alongwith TGA visible in Echo.

In all the 5cases of tetralogy of Fallot RVH was found. RVH was also found in Echo confirmed cases of Ebstein's anomaly and hypoplastic, left heart syndrome. Tricuspid atresia was found on echo in 1 case which was having LVH and normal ECG was found with systemic anomalous venous drainage, which was also diagnosed by echocardiography.

Table - XI

Correlation of clinical and ECG findings in patients of Cyanotic heart disease

Clinical diagnosis	Number of cases				
	Total no.	With RVH	With LVH	With BVH	Normal
1. TGA (3cases)					
a) Simple TGA	2	2	-	-	-
b) TGA with PS	1	-	1	-	-
2. Tetralogy of Fallot	5	5	-	-	-
3. Tricuspid atresia	1	-	1	-	-
4. Ebstein's anomaly	1	1	-	-	-
5. Hypoplastic left heart syndrome	1	1	-	-	-
6. Systemic anomalous venous drainage	1	-	-	-	1

Different radiological features of congenital heart disease are shown in table XII.

Table - XII

Radiological findings in congenital heart disease

Radiological findings	Number of cases	
	Acyanotic	Cyanotic
Increased pulmonary blood flow	22	4
Decreased pulmonary blood flow	1	7
Normal pulmonary blood flow	27	1
Cardiomegaly	31	7
Dextrocardia	3	-
Shape of heart -		
. Boot shaped	3	-
. Egg on side	1	-
Right aortic arch	1	-
Prominent pulmonary artery	12	-

Increased circulation was found in 21 cases of cyanotic heart disease (ASD,VSD and PDA) while 28 cases had normal pulmonary circulation. Among the cyanotics 4 cases were having increased circulation (TGA and hypoplastic left heart syndrome), one had normal and 7 had oligaemic lung fields (case of Tetralogy of fallot, Tricuspid atresia and Ebstein anomaly).

Cardiomegaly was found in 38 (61.3%) cases out of all 62 cases. Thus 31 cases (62%) of acyanotic patients and 7 cases (58.3%) of cyanotic patient had cardiomegaly.

Dextrocardia was found in 3 patients, Boot shaped heart in 3 cases of tetralogy of fallot out of 5 and Egg on side shaped heart was seen in only one patient of TGA out of 3. In a single case, right aortic arch was found.

Prominent pulmonary artery was found 12 cases among them 3 were of PDA, 6 were of VSD, 2 were of ASD and VSD both and 1 was having ASD.

Correlation of radiological findings according to individual acyanotic heart lesions is shown in table XIII.

Table - XIII

Correlation of clinical and radiographic findings in patients of Acyanotic heart disease

Clinical Diagnosis	Number of cases			
	Total no.	Pulmonary blood flow		
		Increased	Decreased	Normal
1. VSD (30 cases)				
a) Simple VSD	23	11	-	12
b) VSD with PS	4	-	1	3
c) VSD with pulmonary hypertension	3	3	-	-
2. ASD (8 cases)				
a) Simple ASD	4	3	-	1
b) ASD with PS	1	-	-	1
c) ASD + VSD	3	1	-	2
3. PDA (5 cases)				
a) Simple PDA	2	1	-	1
b) PDA + Pulmonary hypertension	1	1	-	-
c) PDA + VSD	2	2	-	-
4. Endocardial cushion defect	1	-	-	1
5. Coaretation of aorta	1	-	-	1
6. Aortic stenosis	1	-	-	1
7. Aortic and mitral insufficiency	1	-	-	1
8. Pulmonic stenosis	1	-	-	1

Among 23 cases of simple VSD, 12 had normal pulmonary circulation and another 11 had increased pulmonary blood flow, when pulmonary stenosis was associated with it and found in 4 case one case had decreased pulmonary flow and 3 had normal circulation. Pulmonary hypertension was present in 3 cases of VSD, both of them had increased pulmonary blood flow.

In case of ASD, increased pulmonary flow was found in 3 and normal in 1 case out of all simple cases of ASD.

In all the 3 cases of ASD with VSD radiological picture was different. The pulmonary blood flow was found normal in 2 and increased in 1.

In cases of simple PDA out of 2, one had normal and another had increased pulmonary circulation. There were 2 cases of PDA with VSD and both of them had increased pulmonary blood flow.

In all other types of acyanotic heart disease as endocardial cushion defect, aortic stenosis or insufficiency, coarctation of aorta and pulmonary stenosis pulmonary circulation was found to be normal.

Table XIV shows the pulmonary blood flow in patients cyanotic heart disease.

Table - XIV

Correlation of clinical and radiographic findings in patients of cyanotic heart disease

Clinical diagnosis	Number of cases			
	Total no.	Pulmonary blood flow		
		Increased	Decreased	Normal
1. TGA (3 cases)				
a) Simple TGA	2	2	-	-
b) TGA with PS	1	1	-	-
2. Tetralogy of Fallot	5	-	5	-
3. Tricuspid atresia	1	-	1	-
4. Ebstein's anomaly	1	-	1	-
5. Hypoplastic left heart syndrome	1	1	-	-
6. Systemic anomolous venous drainage	1	-	-	1

The pulmonary flow was found to be increased in all the 3 cases of TGA whether alone or associated with pulmonary stenosis.

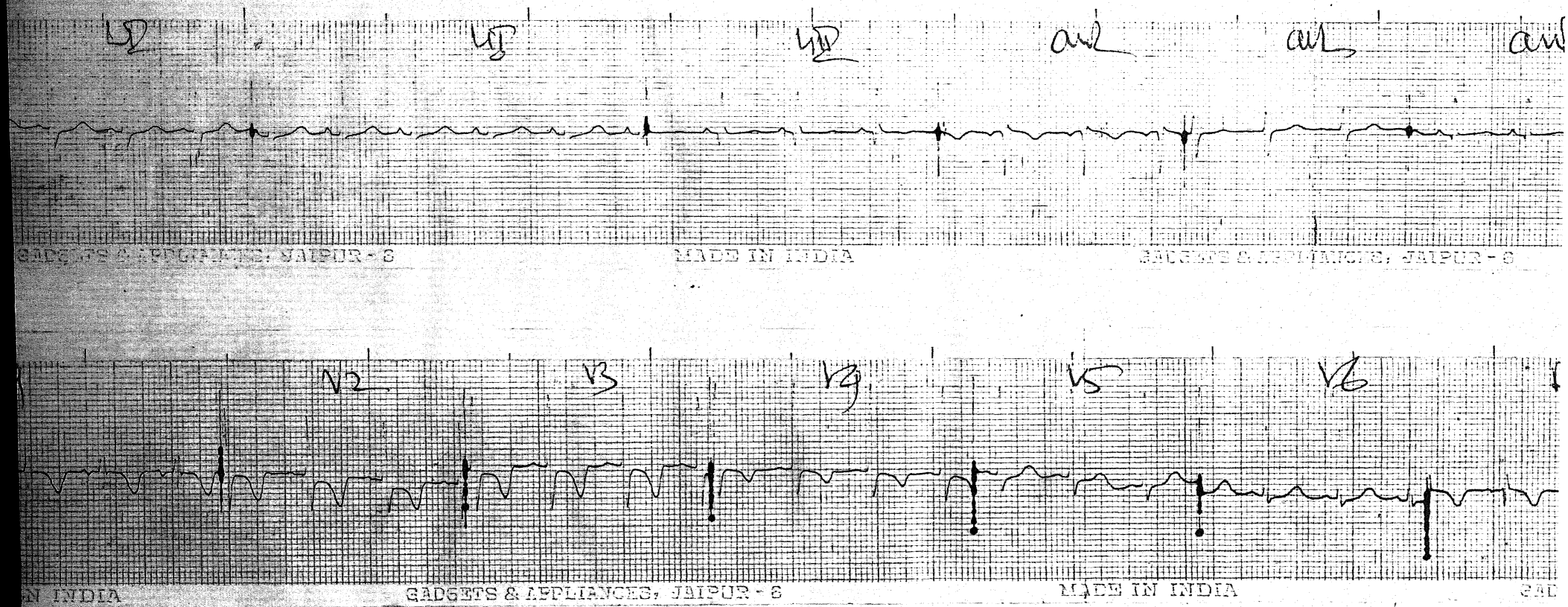
All the 5 cases of tetralogy of Fallot had oligemic lung fields. Oligemic lung fields were also found in cases of tricuspid atresia and Ebstein's anomaly.

Normal pulmonary blood flow was found in the case of systemic anomolous venous drainage. In case of hypoplastic left heart syndrome, the pulmonary circulation was increased.

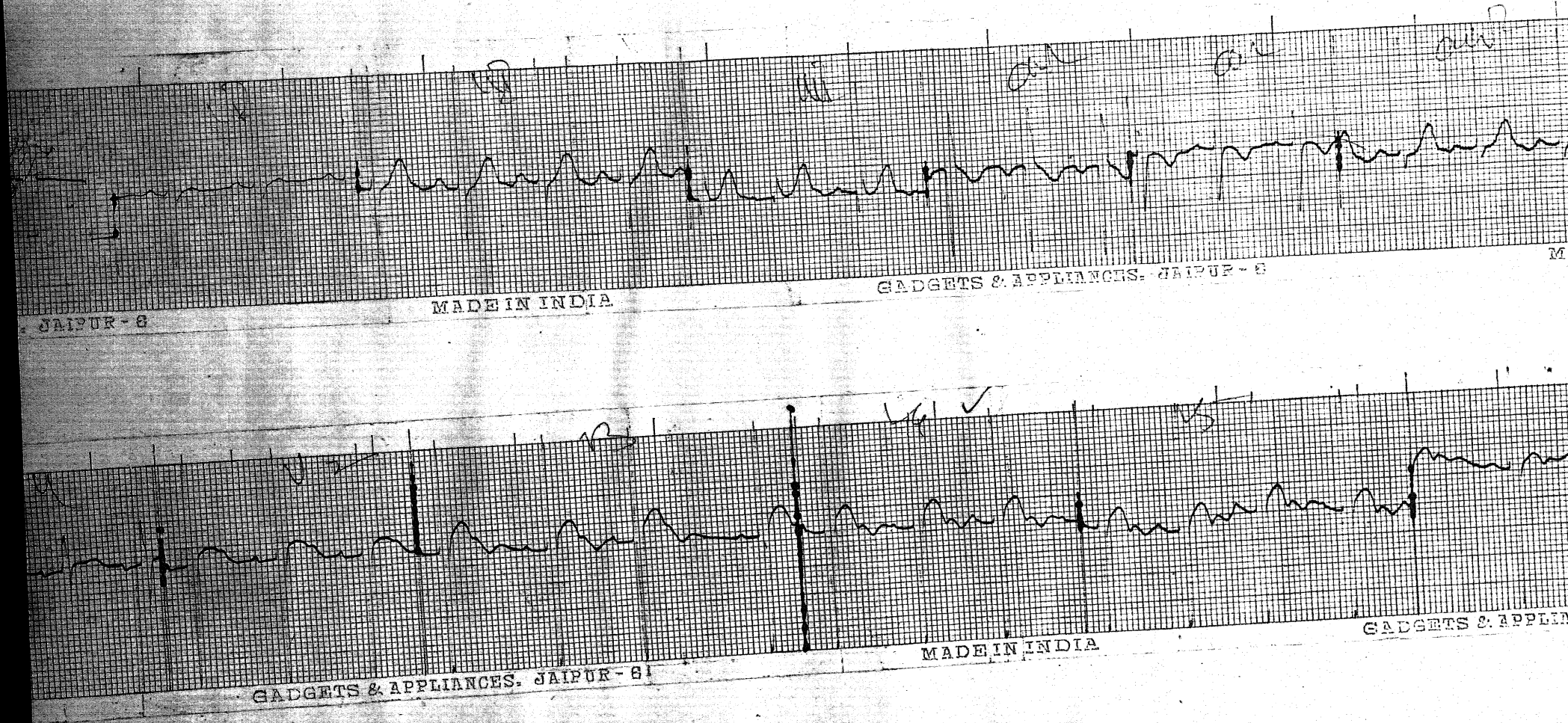
Echocardiographic finding of 40 patients out of 62 were also taken into consideration, when they were followed up to confirm our diagnosis. In 44 (70.9%) cases the diagnosis was made by clinical examination and later confirmed by ECG, X-ray in all and echocardiography in 25 cases. In 10 cases, diagnosis was improved by the help of X-ray and ECG and later confirmed by echocardiography. But in 8 cases, diagnosis could be made only after echocardiography.

..*.*.*.*.*

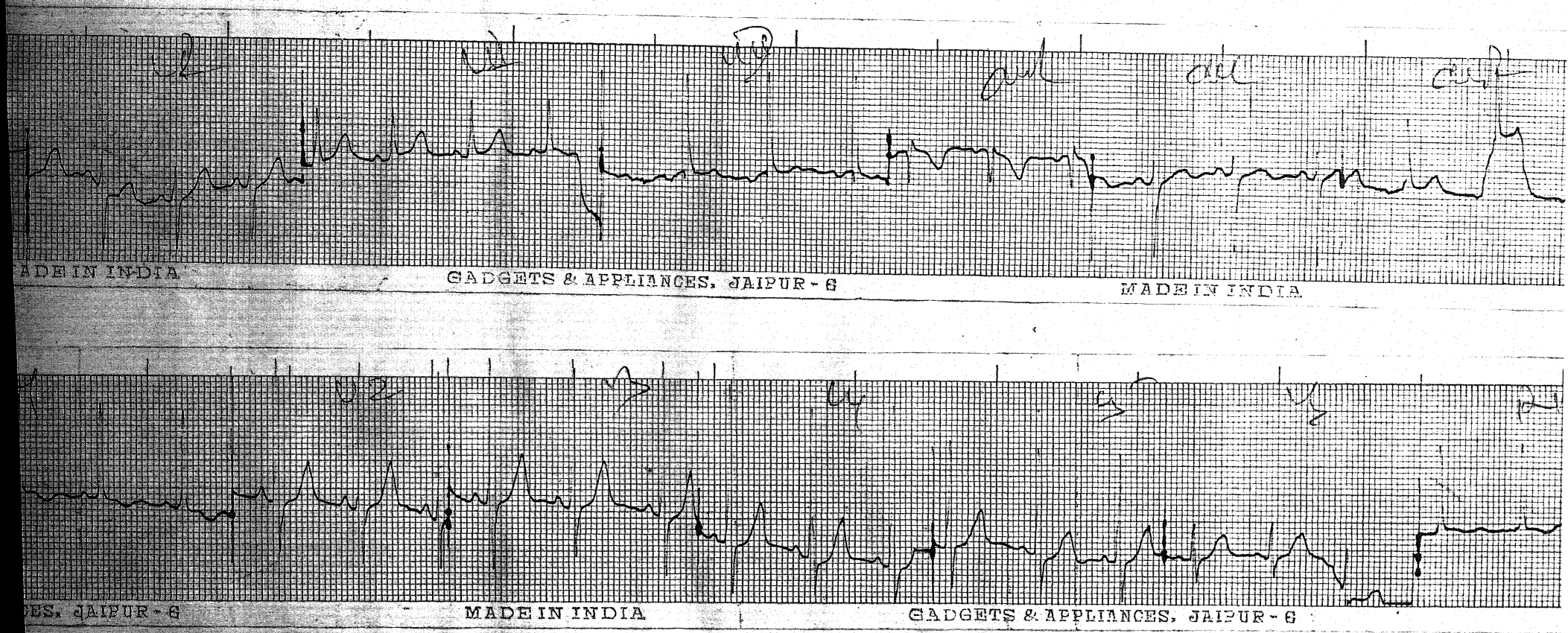
ECG of 3 Yrs. old patient of ASD showing R.B.B.B.



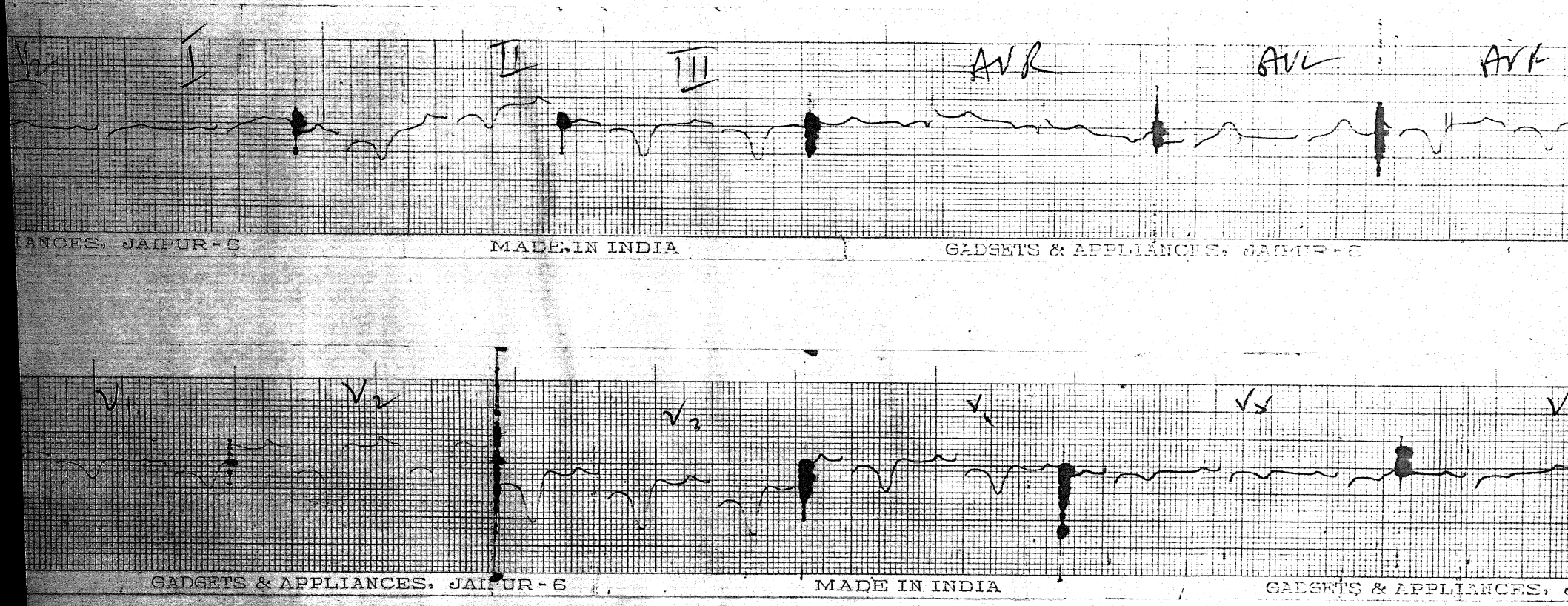
E.C.G. of 1.5 yrs. old patient of V.S.D. showing B.V.H.



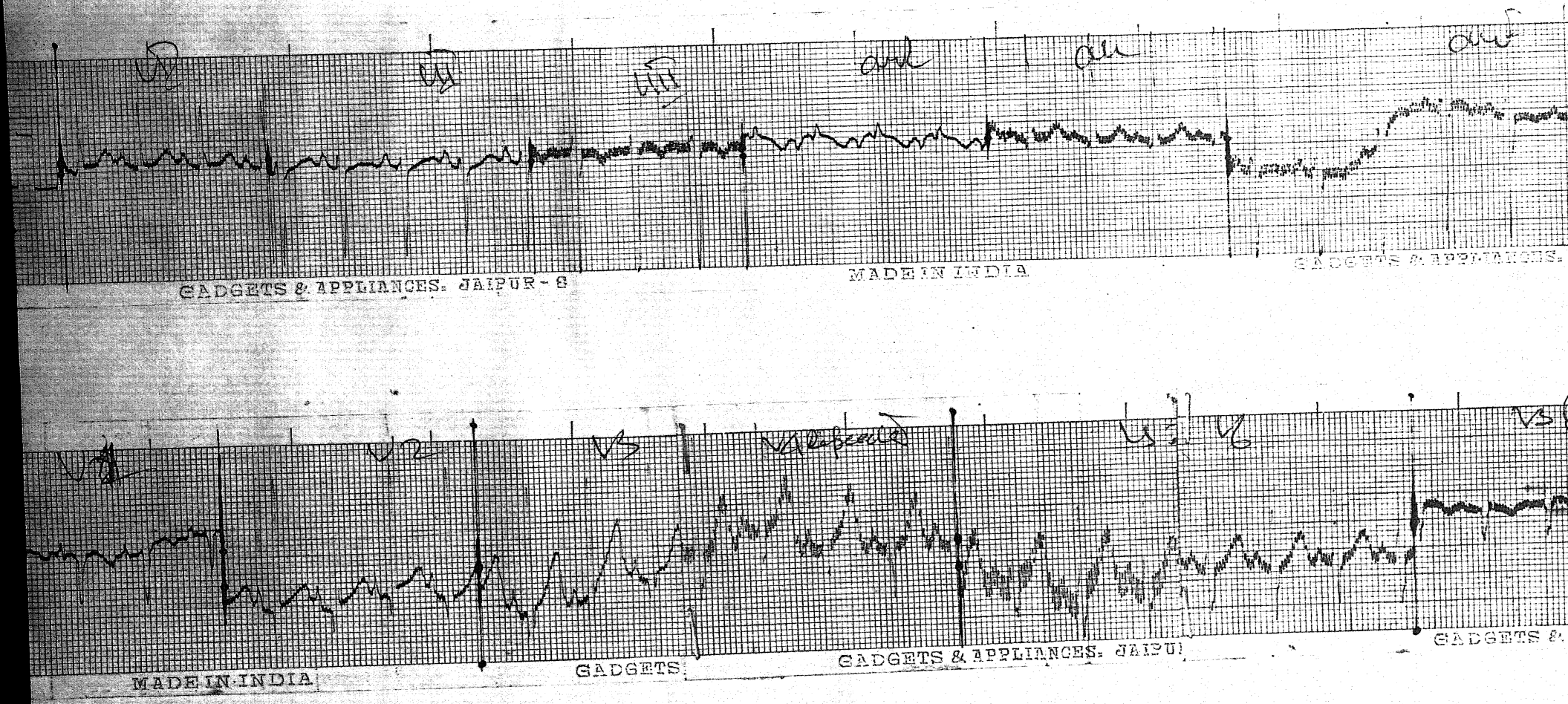
E.C.G. of 5 yrs. old patient of Tetrology of Fallot showing RVH



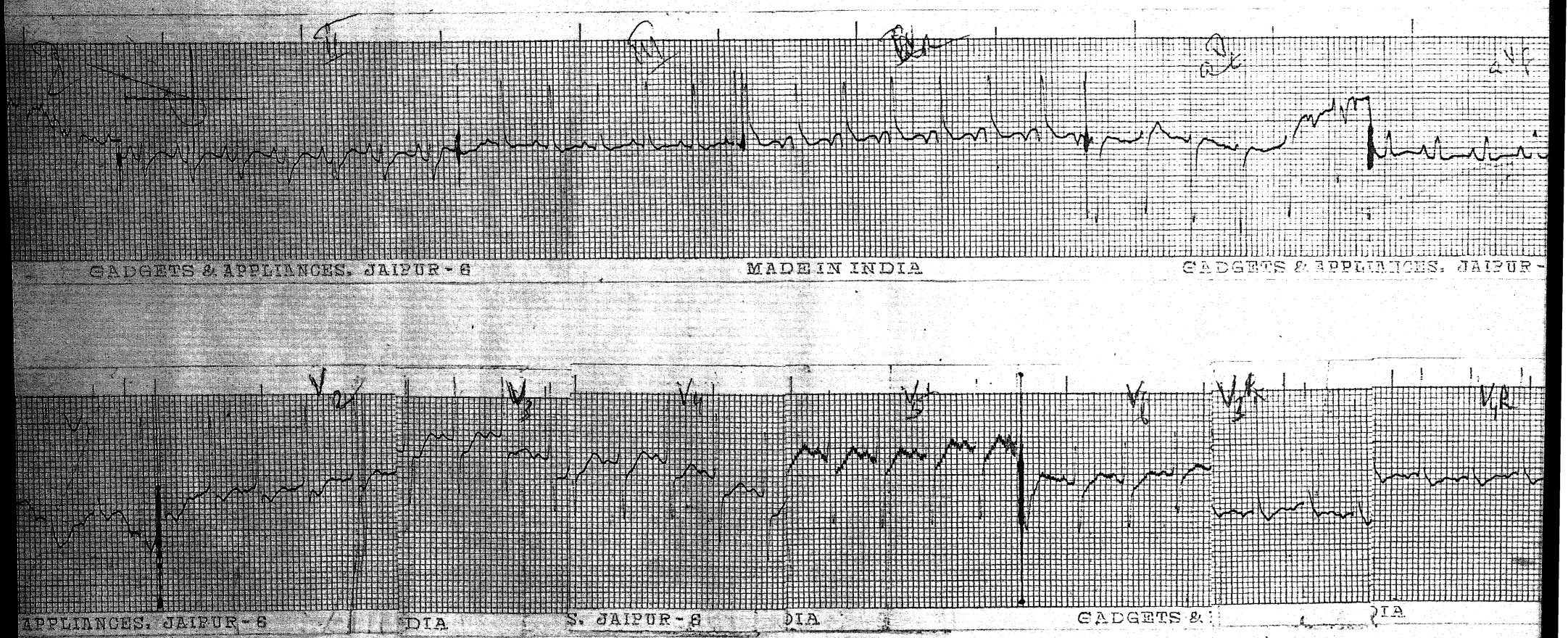
E.C.G. of 13 yrs. old patient of Pulmonary stenosis showing R.V.H. with strain pattern.



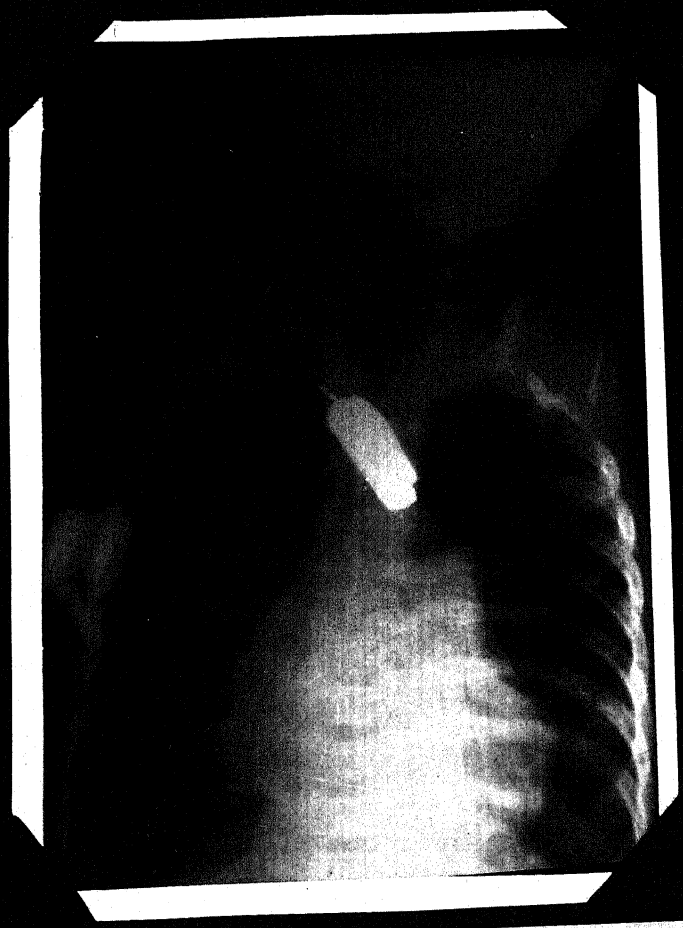
E.C.G. of 1 month old patient of T.G.A. with P.S. showing 'P' Pulmonale, L.V.H. and left axis deviation.



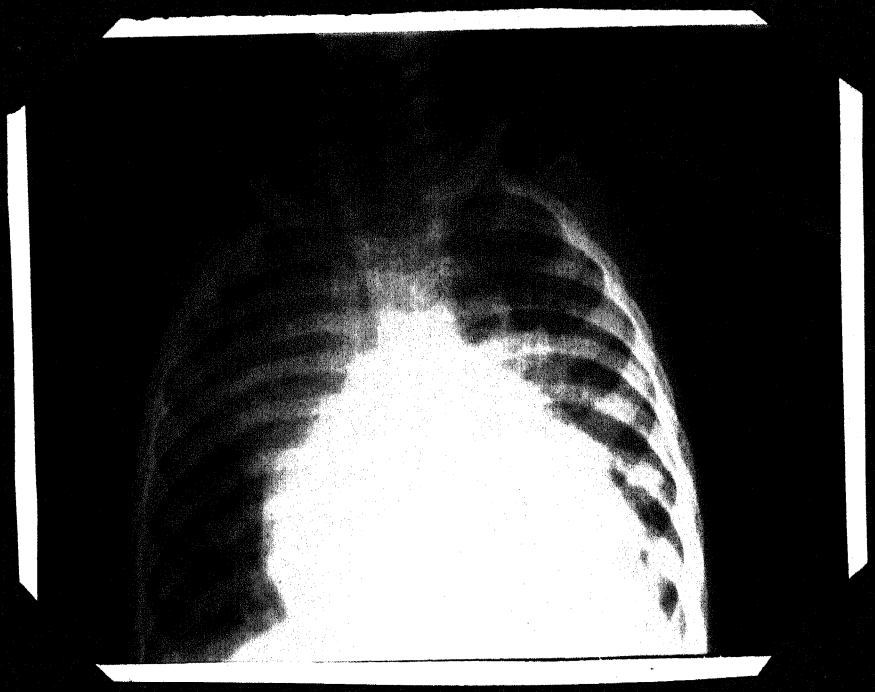
E.C.G. of 8 days old patient of Hypoplastic left heart syndrome showing R.V.H. with marked right axis deviation.



X-Ray chest PA view, gross cardiomegaly in a case of V.S.D.

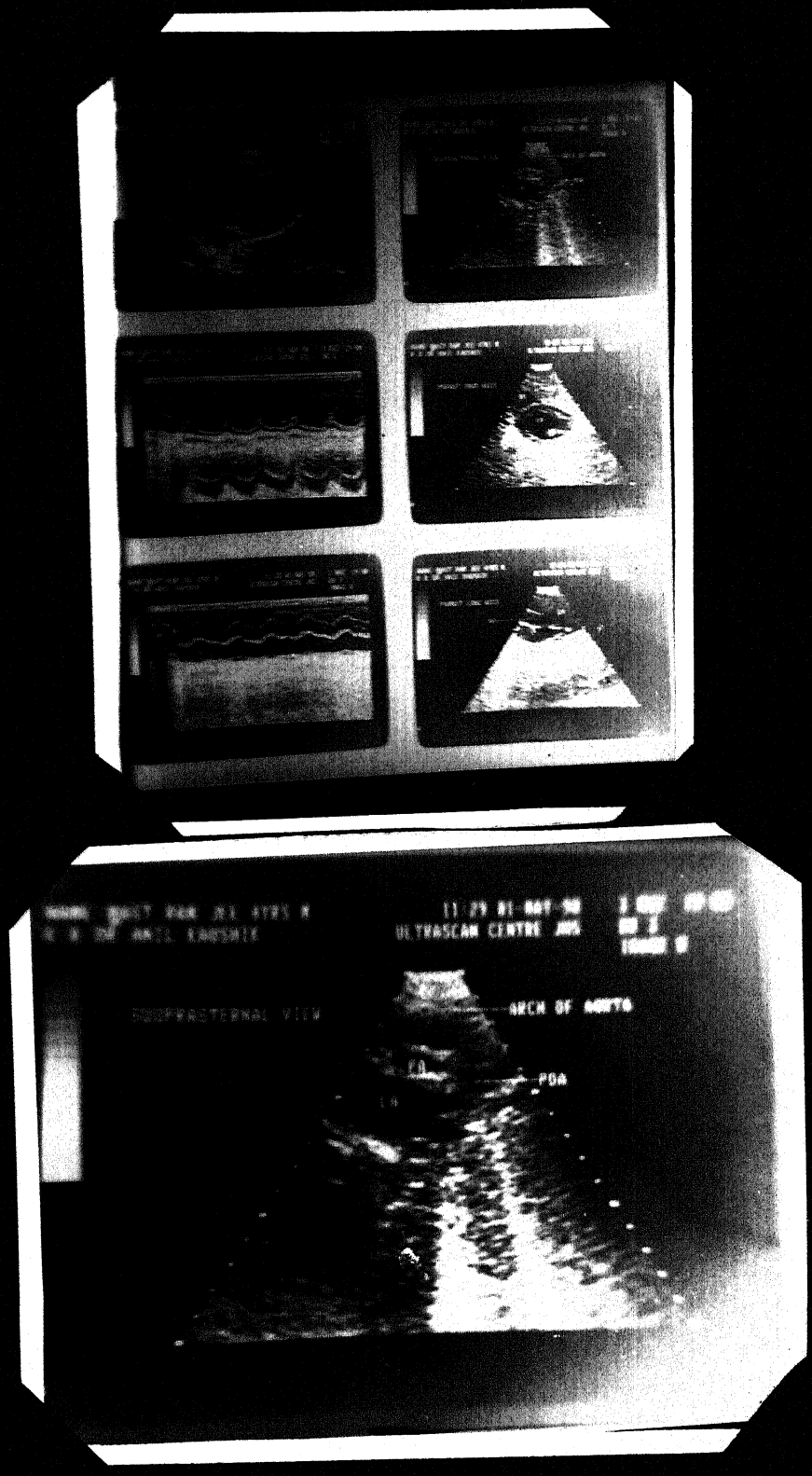


X-Ray chest PA view, prominant pulmonary conus in a case of A.S.D.

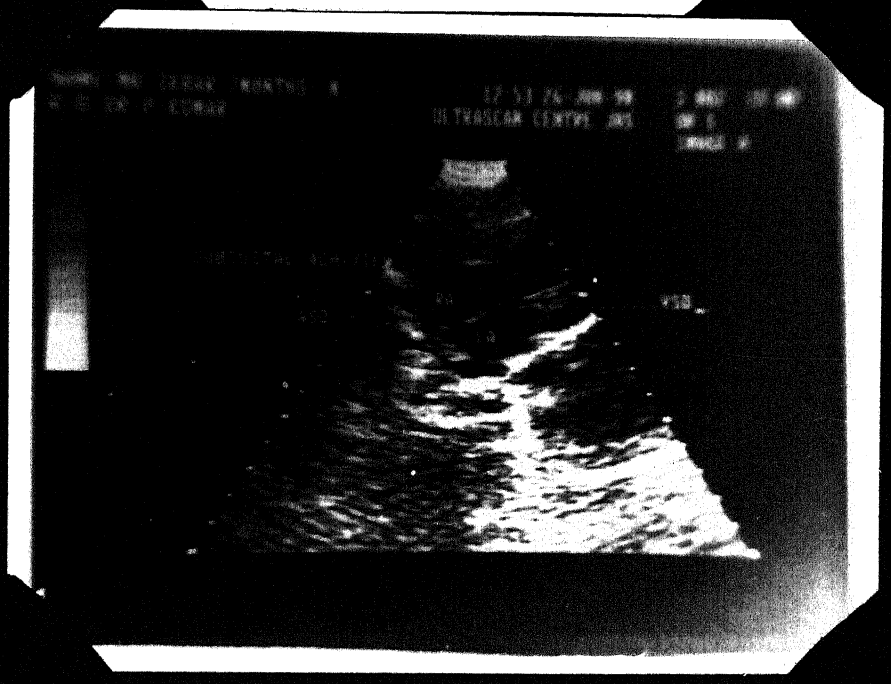


X-Ray chest PA view, Egg on shaped heart with pleuthora, in a case of T.G.A.

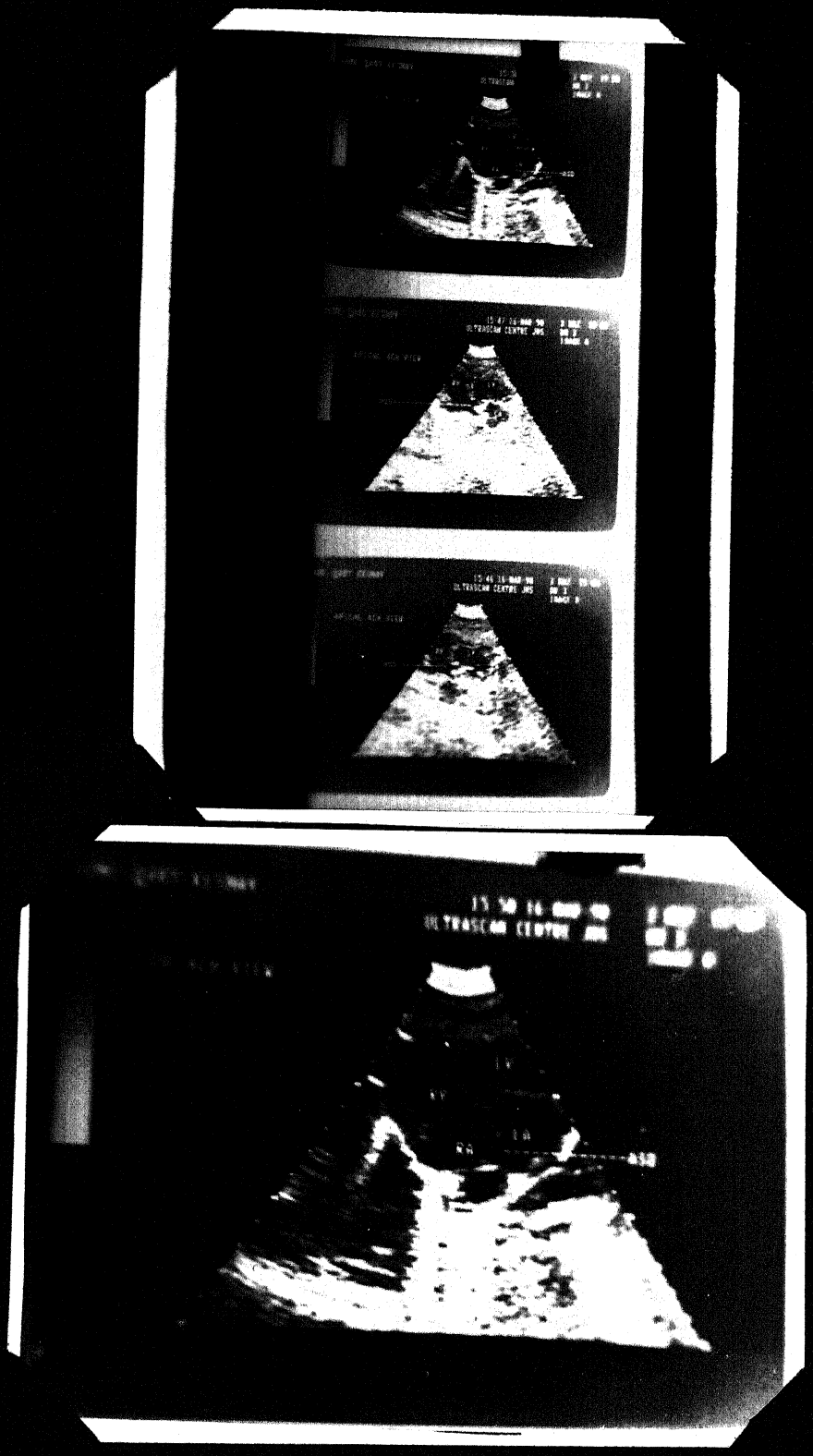
X-Ray chest PA view, BOOT shaped heart with oligemic lung field.



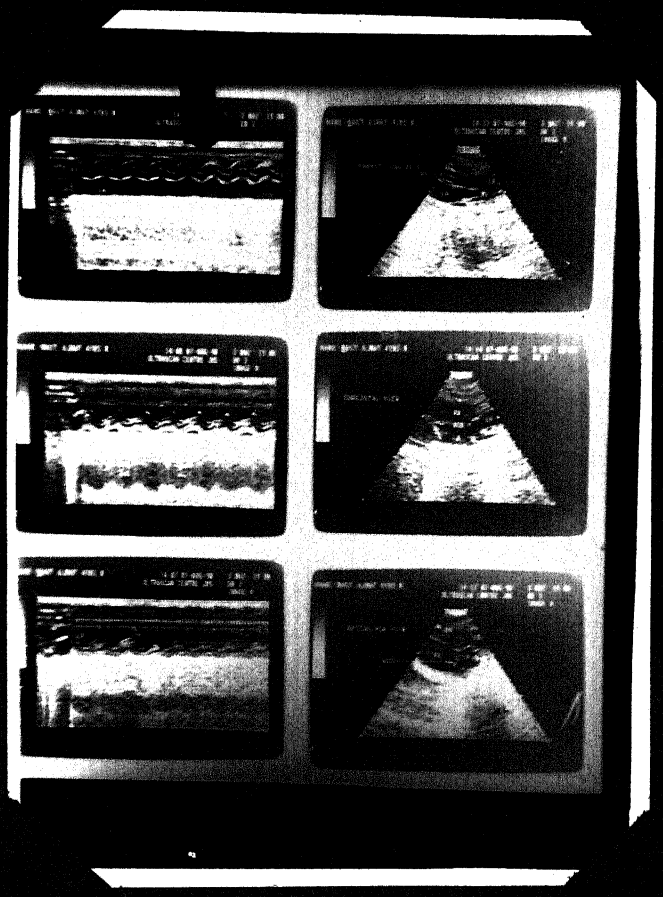
Echocardiogram in a case of P.D.A. showing enlarged LA and LV with P.D.A.



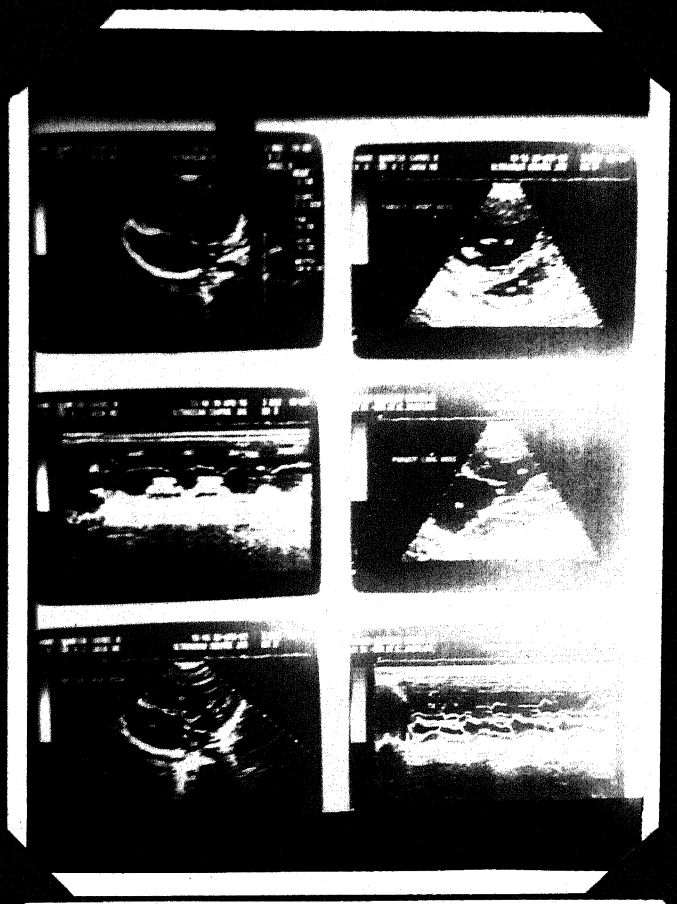
Echocardiogram in case of A.S.D. with V.S.D.



Echocardiogram of a case of A.S.D. with V.S.D.



Echocardiogram in case of A.S.D.



Echocardiogram in case of congenital pulmonary stenosis showing enlarged RA & RV, RVH and prominant 'A' wave in M.Mode Echo.

Echocardiogram in case of Hypoplastic left heart syndrome, showing smallLA & LV with enlarged RA & RV.

DISCUSSION

DISCUSSION

The present study dagcribes an effort to have a surveillance for congenital heart diseases. This study has been carried out on patients of various types of congenital heart diseases, who attended the out patient department of Pediatrics or admitted in Pediatric ward of M.L.B. Medical College Hospital, Jhansi from September 1989 to August, 1990.

As per outdoor patient records of Pediatrics department, the prevalence of congenital heart disease was found to be 1.45%. Higher prevalence was reported by Padmavati and Datey (1968) from various Indian city hospitals as 4.8% in Delhi (1952-1956), 2.3% in Madras (1946), 3.6% in Amritsar (1953), 6.3% in Bombay (1952-1956) and 1.6% in Lucknow (1953). Banerjee et al found it to be 7.5% in all autopsy study from 1964-1975 at P.G.I., Chandigarh. Such data were mostly from large referral hospitals, so there, it is likely to be some concentration of cardiac cases. Moreover, their figures were of patients of all age groups both pediatric as well as adults. Neverthless, we admit some dilution of data in our study due to undiagnosed cases in the register. Various other studies are also available from different countries, where incidence of congenital heart disease was calculated at the

time of birth varied from 0.7% to 1.17% of live births as reported by Neel (Japan) 1948-1954, Richards et al (New York) 1946-1953, Kerebijn (Holland) 1958, Yerushalmy (California) 1970, Mitchell et al (U.S.A.) 1971 and Rose (Canada) 1971.

At birth, diagnosis of congenital heart disease is difficult. Symptomatology of various congenital heart disease appears clearly at later age, moreover, benign murmers are commonly heard at birth.

In order to know the incidence of various types of congenital heart diseases we classified our patients according to the classification given by Beverly, C. Morgan (1978). Acyanotic patients constituted the bulk i.e. 80.6% and the cyanotics were only 19.4%.

V.S.D. had been the most common type of congenital heart disease so far been observed by various workers. We also found 48.2% of our patients having V.S.D. while Keith et al (1974) reported it to be 28.3% in hospital of sick children, Toronto from 1950 to 1973, Krovetz et al (Gainesville) 26%, Gassel (Chicago) 18%, NERICP 16.6% and Srivastava and Tondon (AIIMS, Delhi) 27%. The higher values in our set up can be because it is an easily diagnoseable condition, and it is likely to be easily picked up for referral by practitioners of poorly equipped peripheral areas of Jhansi because of its loud murmer and thrill.

Roger in 1879 described few asymptomatic patients with cardiac findings similar to that of VSD. Since than all asymptomatic small ventricular septal defects have been called Roger's disease. We noted 26.7% of our patients having small VSD, a finding comparable to 24.5% of Nadas and Fyler's study. Keith reported it in 40% of his VSD cases. There is likely to be some dilution in our data as patients of this category are usually asymptomatic or have mild symptoms, they are usually not taken to hospital in our set up contrary to the routine regular check ups of children in developed countries.

Fifty percent of our patients of VSD fall in group having clinically large type of VSD. Its incidence reported by other workers were 82% (Wood) 36% (Bloomfield) and 50% (Nadas and Fyler).

The second common anomaly in our study was ASD comprising 12.9% of all patients. This is very near to various previous studies - 10.3% (Keith et al. 1974), 8% (Gassel) 8% (Krovetz et al)and 13% (Srivastava and Tondon) NERICP study however reported comparatively lower incidence i.e. 2.9%. But this study was conducted in infants and at this age ASD patients usually remain asymptomatic and many may remain undetected unless revealed in routine check up. which is a rarity in our set up.

Next in the occurrence was PDA, as 8.1% of our patients exhibited this type of defect. The figures from other workers ranged from 6.1% to 11% (Keith et al, 9.8%, Krovetz et al 11%, Gassel 11%, Srivastava et al 11% and NERICP 6.1%).

Five (8.1%) patients from present study had tetralogy of Fallot. Keith et al observed 9.7%, Krovetz et al 11%, NERICP 8.9%, Gassel 13%, Paulwood 11% and Srivastava et al 17% of their patients. Our values are slightly lower than others but it will not be wise to compare them due to small sample size of our study.

Next in occurrence was transposition of great arteries, as 4.9% of our patients had it. Previous studies reported it to range between 4% to 5% by various workers (Keith et al, 4.9%, Gassel 4%, Krovetz et al 5%).

Male dominance in patients of congenital heart disease is a universally known fact. The male female ratio in our study was 1.9 : 1. Banerjee et al (1975) also found it to be 1.7 : 1 in P.G.I., Chandigarh a figure very close to that recorded in the present study Muir, Carlgren, MacMahon et al, Gardiner & Keith, Abbott, Roberts, Campbell and also in NERICP studies, male preponderance was noted. Thus it follow the general rule of more congenital anomalies of all types to be more common

in males. Exception to this was PDA, where we found more girls than boys (40%). The female dominance in PDA was also noted by MacMahon et al (60%), Paulwood (70%), Keith et al (69%) and Campbell (73%).

VSD, Tetralogy of Fallot and transposition of vessels had higher male incidence of 70%, 100% and 67% respectively in our study. The male incidence for VSD was recorded to be 59% by MacMahon et al; for Tetralogy of Fallot were 61% (MacMahon et al) 60% (Keith et al) 64% (Paulwood) 59% (Campbell) and for TGA were 73% (MacMahon et al) 67% (Paulwood) and 68% (Keith et al). It could be by chance that all five patients of tetralogy in our study were male.

In case of ASD, we had equal number of children of both sexes. MacMahon et al also found similar results, but Keith et al and Campbell noted relatively more members of females than males i.e. 60% and 66% respectively.

In other groups of heart diseases, we can not comment upon sex ratio as there were very few patients in each group.

The incidence of other associated congenital defects are much higher in children with congenital heart disease than in general population (MacMahon et al, 1952, Campbell, 1965). Various figures from earlier studies varied from

16% to 28% i.e. (28% from NERICP studies 1968-1977, 25% by Greenwood et al, 1975, 21% by MacMahon et al, 1974 and 16% by Lamy et al, 1957). Surprisingly, the incidence of associated anomalies with congenital heart disease was low in our study i.e. 6.4%. Some chances of missing various internal abnormalities might be there as no specific investigation for different systems were undertaken except for heart.

Family history of congenital heart disease was present in 3 cases (4.8%). Among them one had same lesion as that of the patient i.e. VSD. Mckeown et al (1953) found it to be 1.8% , Campbell (1965) 1.7% and Fuhrman (1961) 2.7%.

Several previous studies which linked Oestrogen/ Progesterone to congenital heart defects reported approximately twice the expected prevalence of heart defects among infants with prenatal exposure to exogenous female sex hormones in early pregnancy (Harlap et al, 1977, Nora et al 1976, Heinonen et al, 1977). Harlap et al, 1977 observed the risk of major malformations as major CNS, GIT or genitourinary malformation, cleft lip/palate, polydactyly, syndactyly, hip dislocation, heart diseases, Down syndrome, Glucose-6-phosphate dehydrogenase diffiemby to be 26% higher in group exposed to nonexposed ones.

whereas for minor malformations as hypospadias, Inguinal/ or umblical hernia, hydrocele, haemangiomas and Telangi-actasia, the increase is about 33%. Part of this increase in risk may be due to tetratogenic effects of these hormones.

Janerich et al (1977) reported strong association with prevalence ratio of 6.5. Some positive association was also noted by Rothman et al, 1979. In our study, in 6.5% of patients, a positive history of Oestrogen/Progesterone intake was present.

History of taking antitubercular drugs in 2 of our cases can be a coincidental finding as no positive association to congenital heart defects is available in literature except for phenobarbitone and phenothiazine (Rothman et al, 1979, Heinonen et al, 1977).

In a single patient, history of overexposure to radiation during fourth month of gestation was available. This association can be either by chance or may have some importance as Cox (1964) noticed that all the malformations in general were twice as common in children of mothers, who had been exposed to frequent X-ray examination during gestation.

The most common presenting symptoms of acyanotic heart diseases were cough and breathlessness (46%) recurrent chest infection (36%), feeding difficulty (30%), failure to thrive (24%) and exertional dyspnoea (12%).

Chances of recurrent chest infection are more due to increased pulmonary circulation because of left to right shunts. Apart from this cough can also occur due to congestion of abdominal viscera from right heart failure, which can also cause dyspeptic symptoms as diarrhoea and vomiting (Friedman, 1984). Failure to thrive is a consequence of decreased systemic output, congestive heart failure leading to feeding difficulty and due to negative balance caused by recurrent chest infections.

Cyanotic heart disease patients presented with exertional dyspnoea (67%), cough and breathlessness (50%), cyanotic spells (41.6%), squatting (25%), feeding difficulty (29%) and failure to thrive (41.6%) in this study. Exertional dyspnoea occurs due to arterial unsaturation especially following exercise which increase oxygen demand. But this actually decreases on exertion because of increased right to left shunt, thus chemoreceptors get stimulated causing dyspnoea. An equally common cause of dyspnoea is due to increased pulmonary blood volume and increased pulmonary capillary pressure inherent in left sided failure. Cyanotic spells are said to be expression of cerebral anoxia resulting from drop in arterial saturation due to sudden spasm or right ventricular infundibulum resulting in precipitous drop in the pulmonary flow which usually follow a severe exertion. Franch (1963) described an other possible mechanism

presupposing that any stimulus that decreases blood and tissue PO_2 and pH and/or raises the PCO_2 causes hyperpnea, which in turn increases systemic venous return. Since pulmonary blood flow is fixed or decreased, an increased volume of poorly oxygenated blood is shunted into aorta. Gumtheroth (1965) suggested that hyperventilation with its consequent increase in venous return to the obstructed right ventricle may be one of the underlying cause of hypoxic spells.

However, the severity of these symptoms depends upon degree of the pulmonic stenosis. Squatting, described by Tausing is a characteristic posture assumed after exertion or motionless standing with certain types of congenital heart diseases especially tetralogy of Fallot. This causes exclusion of highly unsaturated lower extremity blood from circulation and augment the peripheral resistance, thus diminish the degree of right to left shunt. Vijay Priya et al (1979) noted presence of cyanotic spells and squatting in one third of their tetralogy patients. In our study 80% patients had cyanotic spells and 60% had history of squatting.

Assessment of Development Quotient (D.Q.) is one of the methods of expression of development, for the purpose of comparison (Prabhakar *et al*). Thus on calculating develop-

mental quotient in each case, we found that there was significant retardation in motor field compared to the social and speech. The delay was more marked in the cyanotic patients. Possible explanations for the growth interference include malnutrition, tissue anoxia, diminished peripheral blood flow, chronic cardiac decompensation, genetic and endocrine factors and frequent upper and lower respiratory infections (Friedman, 1984).

Ruth et al (1982) studied developmental delay both in motor and mental development with congenital heart disease by means of Bayley scales of Infant Development and clinical neurological examinations. The abnormalities can be attributed to decreased arterial oxygen saturation, physical incapacity and psychological and emotional factors.

Feeding difficulties, negative balance and loss of appetite is due to recurrent chest infections and congestive heart failure. Decreased systemic output responsible for underdeveloped muscle mass and decreased physical activity because of that, are the important factors responsible for malnutrition. In cyanotics, unsaturated blood is an additional factor. One of the undernourished child in this study was also having colostomy, done for the high type of imperforate anus. In that case associated malabsorption was an exceptional factor.

As hemodynamic load is small in patients of small and moderate size VSD, these children tend to have a normal ECG. The shunting of blood occurs during systole at a time when right ventricle is also contracting and its volume is decreasing. Therefore, the shunted blood streams to pulmonary artery more or less directly, without any strain to right ventricle. On the contrary, this increased amount of blood passes through lungs and reaches left atrium and then in left ventricle. So when ventricular defects are without pulmonary arterial hypertension the ECG may show left ventricular hypertrophy. Later when either pulmonary stenosis or pulmonary arterial hypertension is developed, they show RVH and LVH or pure RVH.

Welch & Kinney (1942) pointed the association between left to right shunt and pulmonary vascular disease. By later studies also it has been established that by transmission of high pressure from left ventricle to the right ventricle or from aorta to pulmonary artery pulmonary arterial hypertension occurs. In these infants, there is persistance of fetal pattern of lung fields and involution of fetal arterielar changes progress at a slower pace. However, individual susceptibility of arterioles and left atrial hypertension as in VSD also play a role in causation of pulmonary vascular disease. Commonly, it originates from combination of increased flow and increased resistance either singly or both.

Among the 4 cases of pure ASD, normal ECG was found in one and 3 out of 4 had RVH to accomodate and pump large amount of blood. rSR' pattern of right bundle branch block was present in 3 cases in right precordial leads representing delayed posterobasal activation of the ventricular septum. The patient having ASD with pulmonary stenosis had RVH.

In 3 cases both ASD and VSD were present, one of them was having evidence of BVH and remaining 2 of RVH. All these patients presented the picture of large VSD but after echocardiography only, associated ASD was disclosed to us.

Left axis deviation without any evidence of LVH was found in the single case of endocardial cushion defect having common atrio-ventricular canal.

All the cases of tetralogy of Fallot were having RVH. RVH was also found in cases of Ebstein anomaly, hypoplastic left heart syndrome and 2 out of 3 cases of TGA. Evidence of LVH was present in ECG in a case of TGA, a rare finding. Fortunately echocardiography was available with this patient showing pulmonary stenosis significantly obstructing the outlet of left ventricle. There was a single case of tricuspid atresia with LVH on ECG.

Radiological findings of pulmonary plethora were present in most of the cases of large VSD, ASD and PDA due to increased blood flow from left to right shunts. Plethora is a characteristic appearance of the lung vessels found in these condition and rarely from increase in cardiac output in certain other conditions. The main pulmonary artery is enlarged producing a convex pulmonary bay. Among 25% of cases of acyanotic heart disease, prominent pulmonary conus was there, but some cases, it could be concealed by big thymus in children. The pulmonary arteries and veins are increased in size and can be followed into the outer third of the lung. These findings of pulmonary arterial hypertension has to be differentiated from various causes of pulmonary venous hypertension, produced by back pressure to pulmonary veins because of impairment of functions of valves or chambers. In that case distension of normally collapsed upper lobe veins are seen. Later Kerly A and B lines, interstitial oedema, alveolar pulmonary oedema (Bat's wing appearence) and pleural effusion may develop (By Raphael & Donaldson in Textbook of Radiology). Characteristic shape of heart i.e. Boot shaped heart was seen in 3 out of 5 cases, of Fallot's tetralogy in this study. This is due to RVH which lifts up relatively hypoplastic left ventricle and because hypoplastic pulmonary artery. About 28% of patients of Fallot's also have right aortic arch (Pearson & Rigby in Text book of Radiology). We

found it in 20% of cases. Egg shaped cardiac shadow was present in 1 out of 3 cases of TGA which occur because of absence of pulmonary artery at its conventional site, RVH and narrow superior mediastinum. However, in other cases of TGA and Fallots, right ventricular type of cardiomegaly was found.

Out of 50 of the acyanotic group, in 31 patients where echocardiography was possible, diagnosis made out after clinical examination, ECG and X-ray chest remained the same in echo in all of the cases. But then some additional cardiac anomalies were detected, as in 4 cases where we suspected only VSD clinically, both ASD and VSD were seen in 3 cases and in the rest one case, PDA was also present with it.

One case needs elaboration here, which was getting cyanosed while crying and was having signs of ASD. We suspected him to have some cyanotic heart disease, but after echo ASD was found to be present alone without any associated anomaly. The cause of cyanosis was then thought to be right to left shunt occuring during cry. Because of this reason congenital heart diseases are classified in two groups, one having cyanosis and other with little or no cyanosis, as mentioned in Nelson Text book of Pediatrics.

Another case, which was diagnosed as of ASD by clinical examination, ECG, X-ray chest and also by echocardiography, marked clubbing was found on examination and cyanosis was not present at any time. The child was also having imperforate anus for which colostomy was made. So it was interpreted that clubbing in that case was of non-cardiac origin probably due to chronic malabsorption because of long standing (2 years) colostomy.

In cases where small VSD was suspected, echo revealed small VSD in 6 out of 8 cases. Doppler studies are needed for such cases as very small defects can be missed by 2D & M mode echocardiography.

In cyanotic children except for tetralogy of Fallot which could be diagnosed fairly well by clinical examination, ECG and X-ray chest for all other cases echocardiographic studies were found to be useful in diagnosis. For example, a four month old child presented to us with feeding difficulty and cyanosis since birth. ECG showed left atrial overload only and X-ray chest showed Cardiomegaly (left ventricular type). We were no where near the diagnosis. Later, in follow up patient showed the Echo report confirming left atrial and left ventricular dilation. Contrast dye when injected to systemic vein reached in left atrium instead of right and from there to left ventricle and right atrium via ASD. So

a diagnosis of anamolous systemic venous drainage was made, which is a rare condition. In the same patient family history suggestive of some cyanotic heart disease was there in 2 of his siblings.

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SUMMARY & CONCLUSION

SUMMARY AND CONCLUSION

The present study entitled "Study of congenital heart diseases in Bundelkhand region" was performed over 62 patients of congenital heart disease, who attended the Pediatric out patient department of admitted in the Pediatric ward of M.L.B. Medical College, Hospital, Jhansi from September, 1989 to August, 1990. Detailed history of present illness, past illness, family history, antenatal history for exposure of drugs, sex hormones, radiation or any illness dufing that period was taken. Details of developmental mile stones were asked in every case. Detailed physical & systemic examination was done in all the individuals. Routine hematological studies, chest skiagrams and electrocardiogram were done in all the cases. We also included echocardiographic findings of the patients, who already had it with them while they attended the hospital or shown to us in follow up after getting it done at hospital equipped with such facility. 40 out of all had such records.

Diagnosis was made and later they were classified according to classification of congenital heart disease given by Beverly, C. Morgan (1978). Observation were tabulated and later data were analysed.

The prevalence of congenital heart disease was found to be 1.45% as per our hospital records were available. Age of patients ranged from 7 days to 13 years and among them 46.8% were of less than one year of age. Males dominated in the study except for PDA and male female ratio was 1.9 : 1. Family history of congenital heart disease was present in 4.8% patients. Incidence of associated congenital disease was 6.4% in our study. In 6.5% of patients, positive history of Oestrogen/Progesterone intake was present.

There was a statistically significant difference among motor mile stones in both acyanotic and cyanotics while it was insignificant for social and speech mile stones as calculated by observing developmental quotient. 13% patients found to be having marked malnutrition (grade III & IV by Indian Academy of Pediatrics classification) and 87.5% of them (7 out of 8) were having CHF alongwith left to right shunt (6) and TGA (1).

The sample was dominated by acyanotic patients, who constituted the bulk (80.6%) of this series. VSD was the commonest type of anomaly present in 48.2% of our patients. Among them 26.7% were of mild type, 23.3% of moderate and 50% were of severe type of VSD.

ASD comprised 12.9%, PDA 8.1%, Tetralogy of Fallot 8.1% and TGA 4.9% of all the patients. Endocardial cushion defect, Coarctation of aorta, Aortic stenosis, Aortic insufficiency, Pulmonary stenosis, Ebstein's anomaly, Tricuspid atresia, Hypoplastic left heart Syndrome and Anomalous systemic venous drainage were rare anomalies and we found only one case of each group.

Normal ECG was seen in all the cases of mild and 71% cases of moderate type of VSD. But in patients with large VSD, RVH was found in 60% and BVH in rest of them. 3 out of 4 cases of isolated ASD had RVH and rSR' pattern in right chest leads, the remaining had normal ECG.

LVH was the feature of 60% cases of PDA (rest had BVH), coarctation of aorta, aortic and mitral insufficiency. RVH was found in all the cases of tetralogy of fallot, 2 out of 3 cases of TGA, Ebstein anomaly and in hypoplastic left heart syndrome.

Radiologically most of the cases of large VSD, ASD and PDA showed increased pulmonary circulation except the cases which were associated with pulmonary stenosis were lung fields were normal. Oligemic lung fields were seen with all the cases of tetralogy of fallot, tricupid atresia and Ebsteins anomaly. The pulmonary blood flow was found increased in cases of TGA and hypoplastic left heart syndrome.

Cardiomegaly was evident in 62% cases of acyanotic and 58% cases of cyanotic heart disease. Bootshaped heart was seen in 60% case of Fallot's tetralogy while only one out of three cases of TGA had characteristic egg shaped appearance of heart on skiagram. Right aortic arch was seen in only one case of tetralogy of Fallot.

Among 50 acyanotic heart disease patients, in 32 cases echocardiographic report was also available but the diagnosis was not changed which was made by clinical examination, ECG and X-ray. Though it was improved as many combinations or associated defects as ASD and VSD both were seen in 3 of the suspected VSD cases and VSD were also seen in a case of PDAaand bicuspid aortic valve in a case of aortic stenosis were discovered. It also seems to improvise the assessment of size of septal defects. On the other hand among cyanotics, echocardiography helped a lot in diagnosis except for Fallot tetralogy, which was fairly recognised by clinical examination. ECG and X-ray chest.

Following conclusions could be drawn from the present study :

1. Comparable prevalence (1.45%) of congenital heart diseases was found in Bundelkhand region of various type of congenital heart diseases.

2. Males dominated in the study except for PDA in all the types of congenital heart disease. Overall male female ratio was 1.9 : 1.

3. Motor mile stones were significantly delayed in cyanotics compared to acyanotic patients while social and speech mile stones were indifferent.

4. Presence of associated congestive cardiac failure seems to be an important cause of malnutrition in congenital heart disease patients.

5. All acyanotic heart diseases and also tetralogy of Fallot of cyanotic group can fairly be diagnosed by clinical examination, ECG and X-ray chest. In rest of the cyanotic heart disease, echocardiography and other invasive investigations seems to be necessary.

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APPENDIX

DEPARTMENT OF PAEDIATRICS, M.L.B. MEDICAL COLLEGE AND HOSPIRAL, JHANSI

INDIVIDUAL CASE PROFORMA

1. MRD/OPD No.
2. Ward/Bed No.
3. Name of patient
4. Age/Sex M|F
5. Address
6. Chief complaints :
   * cough & Breathlessness
   * Cyanotic spells
   * Failure to thrive
   * Feeding difficulty
   * Excessive sweeting
   * Dyspnoea on exertion
   * Mild exercise intolerance
   * Early fatigue
   * others
7. H/O Past illness :
   * Similar attacks in past
   * Recurrent chest infections
   * Cyanotic spells
   * H/O of joint pain
8. Family history :
8. Antenatal history :
   * Maternal illness
   * Medication
   * Irradiation
10. Geniological history :
11. Developmental history :

    A. Gross motor -

    * Neck holding (3-4 months)
    * Rolls from prone to supine (6months)

* Sitting with support (6-7 months)
* Sitting without support (8 Months)
* Standing with support (9 months)
* Standing without support (10 months)
* Crawling (10 months)
* Creeping (11 months)
* Walking (15 months)
* Climbing up stairs (18 months)
* Running (18 months)
* Goes down stairs (24 months)

D.Q.

B. Manipulative -

* Grasp (5 months)
* Self feeding (6-7 months)
* Transfer the object from one hand to other hand (7Mths)
* Pincer grip (9 months)

D.Q.

12. Social :

* Watches the mother when talked to him (4 Wks.)
* Smile (6-9 Wks.)
* Laughing (3-4 mths.)
* Excites when see toys & bottle (4 mths.)
* Turn head towards sound
* Begin to show likes & dislike of food (6 mths.)
* Responds to name (7 mths.)
* May imitate movements (7 mths.)
* Help in dressing (10 mths.)
* Clap the hands (10 mths.)

13. Speech :

* Monosyllables (7 mths.)
* Combined syllables (8 mths.)
* Jargon (14 mths.)
* Repeat things said (21 mths.)

14. Examinations :

a. General examination -

* General appearance
* Pallor
* Cyanosis
* Clubbing
* Oedema
* Jaundice
* Gen. Lymphadenopathy

b. Examination of C.V.S. -

* pulse
* B.P.
* J.V.P.
* Precordium -
  1. Inspection & Palsation -
  * Apex beat
  * Thrill
  * Other pulsations
  2. Percussion -
  * Rt. Border
  * Lt. border
  * 2nd I.C.S.
  3. Auscultation -
  * Mitral area
  * Pulmonary area
  * Aortic area
  * Tricuspid area
  * others

15. Examination of respiratory system.

16. Examination of C.N.S.

17. Examination of abdomen

18. Anthropometric measurement:

* Weight
* Height/Length
* head circumference

19. Investigations :

* Blood -
  - TLC
  - DLC
  - Hb %
  - ESR
  - PCV
* E.C.G.
* Skiagram chest
  - PA View
  - others
* Echocardiography

0. Diagnosis :

A. Cyanotic

1. Increased pulmonary circulation
2. Decreased pulmonary circulation

B. Acyanotic

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